

Hydrocephalus (3 per 1000)

Def.: Hydrodynamic disturbance of CSF → ↑ vol. Of CSF within ventricular System
with or without concomitant ↑ in its tension.

Types:

- 1- Normal pressure hydrocephalus → only male.
- 2- Communicating hydrocephalus
- 3- Non-communicating hydrocephalus (obstructive) : i most common
- 4- Arrested hydrocephalus

Causes (pathogenesis) :

(A) *Hydrocephalus e` - CSF pressure:*

- 1- ↑ secretion: - choroid plexus (congestion – tumor)
- vit. A (hyper – hypo)
- Idiopathic
- 2- Obstructive : (i commonest)

<i>Congenital</i>	<i>Acquired</i>
A Agenesis of foramen of monro - Arnold chiari type II syndrome	* Post – inflammatory: meningitis. * Post-traumatic * Neoplastic : i commonest post. Fossa tumors
B Bicker Adam's syndrome	
C Congenital toxoplasmosis	
D Dandy walker malformation	

+ **Most common** : X-linked hydrocephalous (aqueductal stenosis)

Malformation of vein of gallen (A.V. fistula – High COP failure – arterial bruit on frontal

& parietal bone auscultation).

3- Impaired absorption:

- * Subarachnoid space: (adhesions – leukemia infiltration).
- * Dural sinus thrombosis.

B- Normal pressure hydrocephalus:

- * Congenital cerebral agenesis.
- * Diffuse cerebral Atherosclerosis.
- * Dementia as Alzheimer.

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C/P

(A) Before closure of ant. Fontanel:

Head	- ↑ head circumference - Delayed closure of a. Font. - Wide separation () sutures - Craniotabes	- Mc Ewen sign :i cracked pot sound on percussion. - Arterial bruit: Malf. of vein of gallen. - Fore shortened occiput: Arnold chairi syndrome - Prominent occiput: dandy walker syndrome
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+ Over stretched & glistening scalp.

Eye:

- * Sun set appearance.
- * Medial squint.
- * Visual disturbance & optic atrophy & papilledema.

Back:

- * Spina bifida.
- * Meningocele

Neurological manifestations :

- * **No - of ICT**

*** Late cases:**

- Mental retardation.
- Motor retardation (spastic paraplegia).
- + ve babinski.
- Convulsions.

b) After closure of A.F.

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- * Head enlargement less significant
- * Manifest of $\uparrow$ ICT
- * Abducent (6th) nerve palsy (medial squint)
- * Personality change

Investigation :**1- X-ray (plain):**

Bfore closure	After closure of A.F.
Cranio-facial disproportion	Signs of $\uparrow$ ICT
Wide speta () sutures	Finger print appearance
Thinning of skull bone	Beaten silver appearance

2- CT & MRI: brain (i best)**3- CSF study :** (cytoalbminous Dissociation)**4- Blood examination.****5- Arteriography****6- U/S :** through ant. Fontanel**7- ECG :** for convulsions.**D.D.: Other causes of macrocephaly:**

- * **Skull causes:** rickets – osteogenesis imperfecta Ch. Hemolytic anemia .
- * **Meningeal causes:** subdural hematome – effusion.
- * **Brain causes :** Hydranencephaly , megaloccephaly (neurofibromatosis).

* **Familial.**

Treatment:

A- Medical:

- * Indication: in cases of $\uparrow$ CSF production.
- * Mech:
 - Salt & H₂O restriction.
 - Diamox.
 - Isosorbide $\rightarrow$ $\uparrow$ CSF absorption.

B- Surgical:

- 1- **Choroid plexectomy:** in papilloma.
- 2- **Diathermy coagulation:** of choroid Plexus.
- 3- **Opening of stenosed aqueduct:** high morbidity.
- 4- **Endoscopic fenestration of 3rd ventricular Floor.**
- 5- **Excision of brain tumor.**
- 6- **Shunt operation:**
 - * I commonest operation.
 - * Using one way valve (spitz – halter valve).

Types	Indications
1- Ventriculo-ureteral 2- Ventriculo-atrial 3- Ventriculo-pleural 4- Ventriculo-peritoneal (most common) 5- Lumbo-peritoneal. 6- Torkilidsen shunt.	1- Progressive head enlargement. 2- Obstructive hydrocephalus. 3- No: - Eye complication - neurological Manifestation - mental affection - ass e` cong. Malformation
Complications: • Obstruction of tube. • Infection . • Rejection. • Shortening of i tube e` age.	

Complications:

<i>Due to hydroceph. Progression</i>	<i>Medical ttt</i>	<i>Surgical ttt</i>
Eye: (Visual dist. – medial squint – papilloedema, optic atrophy – compression on optic chiasma.	- Electrolyte dist. - Metabolic acidosis	1- shunt.... 2- V-P: peritonitis. 3- V-A: endocarditis. 4- L-P: radioculopathy
Neuro: (MR.- UMNL, gait disturb- convulsions – post. Cerebr. art occlusion		

N.B.: CSF.:

* Volume: (adult 120 ml – infant 50 ml).

* Pressure (180-200 mm H₂O = 18 – 20 mm/Hg)

Mental retardation (MR)

Def : Inadequate mental development & social adaptation < 18 yrs.

*Causes:**1- Pre-natal causes:***a- Maternal causes.**

- 1- Placental insufficiency.
- 2- Premature labour.
- 3- Medications
- 4- Poisons.
- 5- Radiation.
- 6- Congenital infections.

b- Fetal causes:

1- Genetic causes:

- * Chromosomal disorders e.g., Down.
- * Familial MR.
- * Congenital hypothyroidism.

2- Inborn errors of metabolism:

- * Phenylketonuria.
- * Galactosemia.

3- Cranial abnormalities:

- * Microcephaly.
- * Craniosynostosis.
- * Hydrocephalus.



2- Natal causes:

- * Birth trauma → I.C. Hge Anoxia & asphyxia.

3- Post-natal causes: **C P K**

- Cerebral trauma.
- Cerebral infections.
- Cerebro – vascular accidents.
- Poisoning e.g., CO
- Post immunization encephalitis.
- Kernicterus.

C/P :

- 1- Developmental → delayed milestones.
- 2- Educational → failure of benefit from – instructns. – experience.
- 3- Social → failure of social maturation.

D: by $I . Q = \text{Mental age} / \text{Chronological age} \times 100$

Grading: - **Mild** : (Moron) → 51 : 75 %

- **Moderate** : (Imbecile) → 21 : 50 %

- **Sever** : < 20 %

Diagnostic criteria:

- 1- IQ. < 75%
- 2- Age of onset of defect < 18 yrs.
- 3- Defective adaptation to surrounding environment.

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Investigations:

- 1- **Karyotype** → in chromosomal abnormalities.
- 2- **Neonatal screening** → in phenylketonuria.
- 3- **X-ray C.T.** → in microcephaly & craniostenosis .
- 4- **Hypothyroidism.**

Mangement:

- 1- **Prophylaxis** → best management. الوقايه خير من العلاج
- 2- **Early diagnosis** → complete cure in some patients "curable MR."
 - a- Cretinism – thyroxin.
 - b- Hydrocephalus – shunt operation.
 - c- Craniosynostosis – craniotomy.
 - d- Phenylketonuria – diet free from it.
 - e- Galactosemia – lactoase free formula.
- 3- **Well established cases** → (ttt according To degree).
 - **Mild degree (Moron)** → Educable & trainable.
Special education needed for optimal develop.
 - **Moderate degree (Imbecile)** → non educable.
Occupational training i only management.
 - **Severe degree (Idiot)** : → Non educable & Non Trainable.
 - **If there is convulsions:** anticonvusant drugs.

Cerebral palsy = static encephalopathy

(i most common motor disorder 2 : 2.5 / 1000)

Def.: clinical syndrome ch` by :

- 3 x yes * Purely motor
- * 2ry musculoskeletal deformity.
- * Clinical manif. appear since birth or early infancy < 3 yrs.
- 3 x no: * Non-progressive lesion.
- * Non-hereditary (not genetic)
- * No mental retardation.
- ± * Special sensory dysfunction (hearing , vision , speech).

A/E:

A- Pre-natal (11%):

- * Maternal: MR.
- * Fetal: - Congenital cerebellar ataxia.
- Fetal anoxia (i most common cause) **MCQ**
- dto: 1- Placenta (infection – premature separation Of membrans)
- 2- Hypotension.

B - Natal: (30 %) : M.R.

C- Post natal: (7 %) : MR.

N.B : Metabolic causes not cause C.P

Clinical types:

1- Spastic (pyramidal) i most common type & may be.

- * Diplegic (L.L.> U.L) – 54 % (**i most common form**)
- * Hemiplegic (one side) – 26 %.

- * Quadriplegic (25%) * paraplegic * triplegic (both L.L. + one arm).
- * Monoplegic (one limb).
- * Manifest: Hypertonia, hyper reflexia , ext. Plant reflex , scissoring of legs , spasticity.

2- Extra A: Choreo-asthetoic C.P.

- * Manifest: hypertonia, rigidity, abnormal movement (chorea, athetosis).

3- Ataxic: lesion in cerebellum – hypotonia, hyporeflexia, ataxia.

4- Mixed : spasticity, athetosis, ataxia.

5- Atonic : floppy infant hypotonia, MS. Weakness, brisk reflexes.

Complications: no sequelae of C.P. (static)

A → attack of epilepsy.

Infection: - lung (bronchopneumonia) most common cause of death
 - Brain : E , M

D → Deformity of bone

Diagnosis :

Clinical diagnosis:

- History of A/E.
- Persistence of Premature reflexes
- Persistence of brain stem reflexes (asymm. Tonic neck reflex).

Invest:

- Assessment of gross motor function.
- Brain imaging (normal in 20% of cases).

Management:

Aim: maximize function & minimize problems.

Types:

- 1- **Physiotherapy:** - Positioning – stretch technique
- Help ms. Relaxation – prevent deformity.
- 2- **Central MS. Relaxants:** diazepam – clonazepam , baclofen – piracetam.
- 3- **Local MS. Relaxant:** (Botulism toxin type A) I most effective.
- 4- **Intrathecal baclofen pumb.**
- 5- **Neuro-surgery.**
- 6- **Hearing aids, speech therapy, educational & training.**

Q. Causes & diagnosis & management of acut conveulsions ???

Causes:a- Non-febrile convulsions:* Toxic:

- Convulsant drugs → aminophylline, steroids.
- Bacterial toxins → tetanus, salmonella.
- Poisoning → CO, lead, organophosphorous
- Post – kernicterus.

* Hypoxic:

- Asphyxia neonatorum.
- Cyanotic spells (cong. Hrt. Dis.)

* Ischemic:

- Cerebral thrombosis (S.C. anemia)
- IC Hg.
- Cerebral embolism.

* Metabolic:

- ↓ Ca - ↓ Mg (tetany + inborn error of metabolism)

* Brain:

- Infection: - Vital: encephalitis, polio enceph.

- Bacterial: meningitis, Br abscess.
- Injury: Trauma
- Edema: AGN.
- Tumors.

N → Nutritional: hypoglycemia – Vit. B6 deficiency.

I → Idiopathic or familial.

H→ICH (trauma, hypoxic ischemic , encephalopathy)

B- febrile convulsions:

- * High environmental temperature.
- * Extra – cranial infection (CVS , Chest , GIT , urinary).

Diagnosis:

1- History: * Onset: (acute – chronic).

* Of i cause: head trauma – infection – cong. Hrt. Dise.

2- Full neurological exam: to decide if cranial or extra-cranial.

3- Investigations:

- * X-ray, CT, MRI.
- * CSF analysis.
- * Blood exam.
- * Urine analysis.
- * ECG, ECHO, EEG.

Treatment:

I- During seizure:

a- Air way: keep patent by:

- * Semiprone position e` head down & to the Rt.
- * Suction of secretions.
- * Mouth gag.
- * Tongue depressor.

b- O2 therapy.

c- Anti-convulsant drugs: IV. Diazepam also, rectal effective (1 / 2 gm / kg).

d- Symptomatic ttt: fever – anti-pyretics acetaminophen + cold fomentation.

e- Underlying cause ttt: infection → antibiotic & hypoglycemia, IV glucose 10 %.

II- Long term management: (Prophylaxis of recurrence)

- **Continuous prophylaxis** : about one year → phenobarbitone.
- **Intermittent prophylaxis** : diazepam at onset of fever.

N.B.: Simple febrile convulsions is benign not has long term sequale – not need prophylaxis.

Febrile convulsions

Def.:

- An event in neurologically healthy infants & children () 6 m- 5 yrs.
- Associated with fever (> 38 C) rectal temp.
- Without (CNS infection – history of febrile seizure).

A/E.: 1- High environmental temp 2- Extracranial infection.

Risk factors:

1- For occurrence of 1st febrile convulsions:

- * + ve family history.
- * ↑ Environmental Temp.
- * Neonatal discharge at age > 28 day.
- * Development delay.

2- For recurrence of febrile convulsions:

- * 1st febrile Convulsion at young age < 15cm.
- * 1st febrile Convulsion has low fever.
- * Family Hx of febrile Convulsion In 1st degree.
- * Family Hx of epilepsy in 1st degree.
- * Many subsequent febrile seizures.
- * 1st complex febrile seizures.

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C/P:

2 forms:

Simple (classic)	Complex (non.classic) = complication
- simple if recurrent not e` in 24 hrs. - no post-ictal phase. - Normal EEG () attacks. - after ↑ Temp - short < 15 min - generalized	- Abnormal age - Recurrent e in 24 Hrs. - post Ictal phase - EEG abnormal () attacks - Abnormal age. - > 15 min - Focal * + ve family history

Floppy infant

Def. : generalized Ms. Weakness.

Manifestations:

1- Frog leg position:

- Legs abducted & flexed on supine position .
- Denotes hypotonia of limbs Ms.

2- Head lag.:

- Head lags backward if pulled from hands in supine position.
- denotes *hypotonia of Neck Ms.*

3- Curved trunk: (ventral suspension)

- Baby suspended in prone Position drops.
- Denotes *hypotonia of trunk Ms.*

A/E.:

1- UMNL:

- Atonic cerebral palsy.
- Cerebellar lesions.
- Chromosomal disorders.
- Metabolic disorders.
- Demyelinating disorders.

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2- LMNL:

A- AHC lesion:

- * **Types :** (acute infantile – juvenile – late childhood).
- * **A/E.:** (Hereditary: Werding Hoffman synd. – acquired polio).

Werding hoffman synd: MCQ

- *I most common cuase of flobby infant.*
- *Manifestation of flobby infant since birth..*
- *Absent grasp Reflex & tendon reflexes.*
- *Worm like movement of tongue.*
- *Pseudo bulbar palsy = late respiratory MS. Paralysis.*
- *Inv. EMG. MS. Biopsy.*

B- Motor neuron disease:

A/E.: - Hereditary

- Acquired: (Guillan Barre synd. Ascending polyneuropathy)

C- Nyoneural junction:

A/E.: - Genetic : Myasthenia gravis.

- Acquired : - Tick paralysis. – Botulism.

D- Myopathy:

- 1- Congenital myopathy → stationary.
- 2- genetic myopathy → MS. Dystrophy always progressive.
- 3- Inflammatory Myopathy (myositis) → regressive (viral, autoimmune).

- 4- Metabolic.
- 5- Endocrinal
- 6- Periodic paralysis.

Hereditary myopathy

Def.: group of progressive hereditary dises characterized By ms. Weakness & atrophy.

Duchene muscular Dystrophy (i commonest)

A/E.: *X- linked recessive* 1/3 cases causes d. to new mutation.

Onset: 1st 5yrs old , after infancy.

Course : progressive – unable to walk at 12 yrs. - death in 1st decade.

C/P.:

1- Weakness of shoulder girdle :

- * Can't raise his above head.
- * + ve slipping sign (slips from exam hands when raised from axilla).
- * Winging of scapula.

2- Weakness of pelvic girdle :

- * Wadding gait.
- * Difficult climbing stars.
- * + ve Gowers sign (climb on himself on getting up).

3- Calf MS.: Pseudohypertrophy.

4- Others: M.R. (mild) + cardiomyopathy.

5- Cause of death: - Resp MS. failure.

- Pneumonia
- Cardiomyopathy.

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INV.:

1- **MS. Biopsy** → D 2- **EMG** → Non specific myopathy 3- **CPK** : ↑

TTT

Contracture : surgical ttt

CHD : Follow up , management

Physiotherapy

Exercise : ↑ Ms power

" *Becker's myopathy* "

A/E.: X-linked recessive

Onset: late childhood.

Course: progressive

Death : At 4th . 5th Decade

" *Cong. MS. Dystrophy* "

A.E.: Autosomal recessive

C/P.: - Floppy infant, no tendon reflex.

- MS. Thin of trunk , limbs.

- Joint contracture.

Prognosis : if sever → fatal

D : MS Biopsy , ↑ CPK

Acute bacterial meningitis

Def.: Acute inflammation of meninges → purulent exudates + neurolog. Manifestation
if not given adequate ttt.

Causative organism :

- Neonate: E-coli & group B-hemolytic strept.
- Infant & child: meningococci Pneumococci , H. influenza (b).

Route: droplet.

I.P : 3-4 days.

C/P.:

- 1- **Acute high grade fever** : (↑ temp) may be normal or subnormal in newborn.
- 2- - **ICT**: 4p + (vomiting – bulging A.F).
- 3- **brain function**: - drowsiness – convulsions. – stupor – coma.
- 4- **Focal neurological signs**: - hemiplegia – cranial nerve palsy.
- 5- **Meningeal irritation Signs.**: neck rigidity , + ve (lasseque sign brudzinski – kernig sign.
- 6- **Skin tash**: petichial generalized in meningococci.

Complication:

- 1- **Septicemia** → purpura fulminans : - Diffuse Hge + Acute addis crisis .
- Death due to DIC (24 hrs).
- 2- **CNS**.
 - Defective motor function
 - Dural effusion (sub).
 - Deafness.
 - Obstr. Hydrocephalus

Investigations:

1- **Blood ex.**: (culture – CBC: ↑ WBC's).2- **CSF**:

	Normal	Acute bacterial	Viral	T.B.
Pressure	< 180	↑↑↑	n. or ↑	↑↑
Aspect	Clear	Turbid	Clear	Opalescence
Protein	20- 40 mg/dl	100-500	40-500	100-500
Glucose	40-80 mg/dl	< 50% of blood	N.	< 50% of blood
Cells	0-5 lymphocytes	100- 6000 PMNS	25-100 lymphocy.	10-50 PMN : lymph.
Bacterial viral	Sterile	+ve Gm stain +ve culture	Virus may be detected	Acid fast bacilli

D.D.:

- 1- **Aseptic meningitis:** sterile CSF. Culture
- 2- **T.B. meningitis:** choroids tubercles + x-ray chest.
- 3- **Brain abscess:** ↑ protein + normal glucose + CT diagnosis.

TTT

1- Hospitalization.

2- I.V. line & fluids & drugs.

3- Lumbar puncture.

4- Supportive ttt:

- ↑ ICT: mannitol IV.
- Shock: fluids I.V.
- Coma : tube feeding.

5- Specific ttt: for 5 days.

- < 6m → ampicillin + garamycin.
- > 6m → ampicillin + chloramphenicol
- Then, acc. To culture & sensitivity Test
meningococci & strept & pneumo → penicillin or ampicillin).
- E-coli & H. influenza → Ampicillin , gentamycin.

* + ttt of complication.

6- Prevention:

- * Chemoprophylaxis: Rifampicin.
- * Vaccination meningococcal Bivalent Vaccine .

من اذكار الصباح والمساء

- سبحان الله وبحمده عدد خلقه ورضا نفسه وزنة عرشه ومداد كلماته .
- بسم الله الذي لا يضر مع اسمه شيء في الارض ولا في السماء وهو السميع العليم .

Haemophilus influenza infection

Causative organism: haemophilus – influenza (6 types a – f) → most virulent is B- serotype.

Mode of transmission : droplet.

IP : variable.

Risk groups: children with:

- Stickle cell anaemia.
- Asplenia.
- Immune deficiency malignancy.

Clinical pictures: may result in:

- 1- Meningitis.
- 2- Cellulitis.
- 3- Epiglottitis.
- 4- Pneumonia.
- 5- Bacteremia.
- 6- Other as: UTI , endophthalmitis, pericarditis, osteomyelitis, otitis media, conjunctivitis or sinusitis.

Diagnosis:

- 1- **CBC** → leucocytosis.
- 2- **Bacteriology:** - Smear stained with gram stain or methylene blue.
- 3- **Culture (chocolate agar)**
- 4- **Serology.**

Treatment:

- 1st line: Augmentin, Azithromycin, 3rd generation cephalosporin.
- Less effective: ampicillin, amoxicillin, chlormaphenicol, Cotrimoxazole

Compare () Human & Cow Milk ?

1- Chemical composition:

	Human milk	Cow milk
1- water	88 %	88 %
2- SP. Gravity	1030 – 1032	1030-1032
3- protein:	1.2%	3.5%
lactalbumin & globulin	60%	15%
casein	40%	85%
4- CHO	7%	4%
5- FAT	4%	4%
6- Minerals (mg %)	1/4 mg/dl	3/4 mg/dl
Na, Cl, Ca, K, P, Mg	↓	↑
Fe	↑ (0.15mg/dl)	↓ (0.1mg/dl).
7- Vitamins		
A,D,C, niacine, riboflavin	↑	↓
Thiamine (B1)	↓	↑

Calories : 67 cal /100 ml.

Fat	50%	50%
CHO	42%	30%
Protein	8%	20%

2- Biological composition:

a- Protein:

* Casein : lactalbumin - 6 : 1 → cow

- 1 : 3/2 → human

* Casein the cause of cured formation so, cured of human milk is more finer , digestible .

* Cow milk cured → boiling, drying, acidification, and alkalization.

b- FAT : of cow : - globules: ↑ (15 times).

-Volatile F.A.: ↓ (7 times).

- Essential F.A. : ↓ so, cause steatorrhea for premature.

c- Minerals : Ca⁺⁺ / P ratio

In human → 1.5 : 1 (optimum for absorption)

In Cow → 6 : 6 (not optimum)

Management of breast feeding ?

1- Initiation of feeding:

* Started within 1st 6 hours (earlier is better).

* Put baby on each breast for 1-3 min.

2- Technique of feeding:

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قبل الرضاعة	تجهيز مرتاحة	Mother sitting in comfortable position
	تنظيفه	Cleaning nipple & areola using fresh water.
وقت الرضاعة	تجهيز الواد	Hold the baby in semi sitting position on mothers arm & holding breast the index & middle 3 fingers of other hand & gentle squeeze by the thumb.

	تستخدم الثديين	Both breasts should be used in the same fed.
		It is advice to exhaust one side before shift to the other, as fat in the last portion of fed.
	توقفه في النص وفي الآخر	At the middle & end of the fed – hold the baby in erect position to allow eructation – to prevent milk regurgitation.
بعد الرضاعة	تنومة على بطنه	After feeding, put the baby in prone position to prevent aspiration if there is regurgitation.
	تغذية	Boiled water allowed to cool can be given at any time.
	أوعى	If supplementary or complementary feeding, never give sweetened milk – the baby likes it & refuse breast.

3- duration of feeding:

- * 1st day → 1-3 min on each breast.
- * 10th day → 5-10 min on each breast.
- * Not allowed → more than 20 min/fed.

4- Frequency of feeding:

- * Two methods:
 - Regular o'clock method: (3 hr interval starts 6am. → 9pm
 - additional mid night fed in 1st 2 months).
- * Demand method:
 - The best method. – Self regulatory.
 - The baby controls his intake physiologically by crying.
- * Frequency ↓ with age:

- 1st 4 months → 3 hr. interval.
- 4-10 months → 4 hr. interval.
- 10-12 months → 5 hr. interval.

Advantages of breast feeding?

Advantages Of composition of breast Feeding ?

Disadvantages of artificial feeding?

إعلان لبن الأم

- 1- Supply necessary nutrients for infant except. (Vit. D, C, fluorine). مغذي
- 2- Easily digestible d. to low cured tension. سهل الهضم
- 3- Cheaper than any form of feeding. رخيص
- 4- Always available in proper temperature. دائما جاهز
- 5- Fresh & free from contam. Bacteria → ↓ risk of GIT distur. نظيف
- 6- More safer as ↓ risk of : * PEM * Rickets * tetany * G.E. يمنع الامراض
- 7- Has antimicrobial action: يمنع العدوي

A- Antibodies: against bact. & viruses (IgA-↑mucosal defense)
(Abs → against E-coli)

B- Proteins: * Vit B12 binding protein → bind vit B12
* Lactoferrin - (bind Fe → ↓ growth of E-coli)
- (Bacteriostatic for E-coli) .

Buffering capacity: of human milk is low so prevent neutralization of stomach Hcl.

Bifidus factor: growth factor for lactobacillus bifidus bacteria
→ harmless & prevent growth of path. bacteria.

C- Cells: granulocytes, macrophages, lymphocytes: phagocytosis.
Complement.

E- Enzymes: Lysozymes, peroxidase.

Interferon: producing cells as T-lymphocytes.

- 8- Has maternal benefits: ↓ risk of postpartum Hge.

مفيد للام

↓ risk of breast cancer.

↓ natural contraceptive method.

9- Psychological adv.: Closest & More relationship satisfaction () mother & baby. **نفسيا**

Stages of human milk secretion ??

1- 1st (2-4) days → colostrum.....

2- 5-20 days → transitional milk.

3- After 21 days → mature breast milk.....من الجدول.

Colostrum

Def.: secretion of I breast in the 1st 2-4 days after delivery

Features:

- 1- Amount: 40-60 ml/day.
- 2- Colour: deep lemon yellow.
- 3- SP.G: high (1040 – 1060).
- 4- PH: alkaline.
- 5- Composition:
 - protein 8% (↑ breast milk)
 - CHO 4% (↓ breast milk)
 - Fat 3% (↓ breast milk)
 - Minerals (↑ breast milk)
 - Calories (58 cal/100ml).
- 6- Microscopically:
 - Colostrum corpuscles
 - Mononuclear, polymorph nuclear leucocytes.
 - Fat globules of variable size.

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Function:

- 1- High nutritive value (↑ protein).
- 2- Contain Ab (IgA).
- 3- Laxative effect: help meconium evacuation.

N.B.: If colostrum persists → breast will not secrete milk.

Difficuties of Br. Feeding & their managment?

A- In the infant:

- 1- *Cong. Anomalies:* * e.g., hare lip - cleft palate - CHD.
* Management: surgical correction.
- 2- *Immatutity & prematurity:* Management: feeding of preterm.....
- 3- *Intracranial Hge:* = Periventricular Hge (conservative ttt)...
- 4- *Mental retardation:* * 1ry or 2ry * Manag: nasogastric feeding.
- 5- *Facial palsy:* * Manag.: nasogastric feeding.
- 6- *Nasal obstruction:* * Bilat. Nasal atresia (Manag: surgical caree)
* Common cold : Manag : 1% epinephrine in saline / 5min
before feeding.
- 7- *Mouth conditions:* * Stomatitis, ulcers, monaliasis
* Manag: medical ttt. Antib., antifungal.

B- In the mother:

Nipple:

- 1- Retracted nipple: Manag : glass or plastic shields for 6 w or more.
- 2- Fissured nipple: Manag : ttt of the cause & antibiotics
- 3- Sore nipple:

<u>Causes:</u>	* Faulty suckling	* Bad antenatal care.
	* Milk not washed after fed.	* Milk engorgement
<u>Manag:</u>	* Stop br. Feed. (1-2 d)	* Local antiblotics.
	* Express milk manually & give it to baby.	

Breast:

4- Abnormal large breast.

5- Acute mastitis & breast abscess: **Manag:**

- * Don't stop breast feed. If no pus in milk.
- * Suitable antibiotic (anti-staph)
- * Express the remaining milk...
- * Incision & drainage .

6- Breast engorgment: milk > infant intake.

Prevention : * On demand feeding.

* Manual expression of part of the milk.

Manag : * Express milk manually.

* TTT of the cause.

* ↓ Milk production (stilbesterol).

Milk:

7- Delayed appearance of milk:

* Occurs after cesarian or shock.

* Manag: Insist on breast feeding & if delayed more
→ complementary feeding.

8- Twin pregnancy: Manag:

1- One breast fed. Other artifical alternatively.

2- If one is weak take breast all feds & the other artificial all feds.

C.I Of Breast. Feeding ?

Permanent	Temporary
Maternal	Maternal
1- <u>Infectious diseases</u>: - Active T.B.: (but may continu under INH) - Syphilis (Not stop Br. Feeding as infection during preg., Delivery) 2- <u>Cachectic disease</u>: - D.M. - Thyrotoxicosis - Severe malnutrition - malignancy - Heart Diseases. 3- <u>Maternal disease</u>: (Neurological) - Epilepsy - sever neurosis 4- <u>Breast</u>: - cancer - sever inverted nipple 5- <u>Pregnancy</u>: weaning is absolute Indication after 20 week.	<u>Nipple</u>: - Fissured - Sore <u>Breast</u> : - mastitis - Breast Abscess <u>Acute illness of the mother</u>: If the infant not have the same infection (septicemia , typhoid , malaria) .
Foetal	Foetal
1- <u>Congenital diseases</u>: - anencephaly - complete cleft palate 2- <u>Metabolic diseases</u>: - galactosemia - phenylkenonuria	1- <u>Mechanical cause</u>: - Severe hare lip. - Feeding by spoon or dropper until surgical correction 2- <u>Acute illness of the baby</u>: - Express milk & give it to him.

حديث قدسى
عن أبى هريره رضى الله عنه قال : قال رسول الله صلى الله عليه وسلم :
" تفتح أبواب الجنة فى كل اثنين وخميس " - قال معمر : وقال غير سهيل : " وتعرض الأعمال فى كل اثنين وخميس - فيغفر للمعز وجل لكل عبد لا يهرك به شيئاً إلا المتهاجرين يقول الله للملائكة : ذرهما حتى يسألما " .
حسن

Efficacy of milk supply ?

1- The look & satisfaction of the baby after feed: if satisfied : sleep 3-4 hrs.

2- Gain in weight: the expected w t : (age + 9 / 2)

* At birth: 3 - 3.5 kg.

* 1st 4 m : 6 kg.

* 2nd 4 m : 8 kg.

* 3rd 4 m : 9 kg.

3- Stool:

* Constipation & small hard → under feeding.

* Diarrhea & bulky offensive → over feeding.

4- Urine:

* Polyuria → over feeding.

* Oliguria → under feeding.

5- Microscopical examination for colostrum corpuscles.:

6- Chemical examination of milk .:

7- Test fed.:

* Weighting infant before & after having breast.

* Normally ↑ wt (2.5 ounce / pound B.Wt /day).

8- +ve milk ejection reflex : good milk supply.

	<i>Under feeding</i>	<i>Over feeding</i>
A/E	* Difficulties of br. Feeding. * Insufficient formula or diluted.	* Irregular feeding time * frequent feeding time * Prolonged // // * Excess formula or concentrated.

<i>C/P</i>	* Crying & not sleep * Suckling his fists. * Fail to gain wt → loss wt. * Constipation. * Oliguria & dehydration. * Test fed. : Below normal. * Pale	* Crying due to colic & flatulence * Vomiting. * Over wt. * Diarrhea. * Polyuria. * Test fed.: Above normal. * Dyspneic & ↑ sweating
<i>TTT</i>	* In breast fed : (TTT the cause – complementary feeding) * In artificially fed: correction of quantity & quality	* In breast fed.: (Feeding time not > 20 min & not frequent. * In artific. fed.: correction of quantity & quality.

Weaning

Def.: Introduction of food other than milk to the infant diet.

Stages:

- a- Beginning of weaning: solid food with breast milk.
- b- Complete weaning: stop breast milk.

Time:

Usually at 6th months or may at 4th m.

Due to after 6th m breast milk not sufficient & poor in iron.

Complete weaning: 1-1.5 yrs (not exceed 2 yrs).

How:

Different from child to child acc. To (appetite – what he likes – what he dislikes).

Rules of weaning:

- 1- Gradually.
- 2- Any new food introduced in small amount (1-2 tea spoonful) , ↑ gradually if likes it.
- 3- New food accepted if thin or diluted.
- 4- New food introduction not > 1-2 times/w.
- 5- New food amount should be left to appetite of the baby.

S/E:

- | | | |
|---|-------------------|-------------------------------|
| 1- Marasmus | 4- Iron ↓ anaemia | 6- Vit D ↓ rickets |
| 2- Gastroenteritis | 5- Colic | 7- Protein induced entropathy |
| 3- Allergic reaction : fish , egg , banana. | | |

Sequence of food introduction:

- **4 m** : milk products (soft cheese – yoghurt) + fruit juice.
- **4 – 6 m** : cereals + potatoes + biscuits + mashed banana.
- **6 m** : egg (start with yolk).
- **7 m** : cooked vegetables.
- **8 m** : (meat + beans).
- **9 - 12 m** : (poultry + fish + other food).

دا مادح & دا البيزي

Feeding program of infant agedmonths ?

1- Expected wt of the baby is: $(\text{age} + 9 / 2)$

2- Nutritional requirements of the baby:

- * Calories: $110 \times \text{kg} = \dots\dots\dots \text{Cal/day}$
- * Water: $150 \times \text{kg} = \dots\dots\dots \text{ml/day}$
- * Protein: $2 \times \text{kg} = \dots\dots\dots \text{Gm/day}$

3- How it can be supplied:

- * Breast milk: duration & frequency.....
- * Artificial milk:

Amount = 150 x kg = ml/day

Frequency: as breast feeding

Type:

- Fresh animal milk
- Dried animal milk.
- Evaporated & condensed milk.

* Mixed:

- Complementary: the fed essentially from breast milk & complemented by other foreign milk.
- Supplementary: one or more feeds s essentially completely substituted by f. milk.

4- **Solid food** : can be given > 6 (4m).... Weaning sequence واشرح ال

5- External additions:

* Vit. A: 2000 Iu/d.	* Vit. B1, B2, B6
* Vit. C: 50 mg/d	* Vit. D: 400-800 Iu/d.
* Fe: 10-15 mg/day	* Zn.

6- Nutritional disorders : which can affect the baby :
(KWO – marasmus – rickets) & its prevention.

Artificial feeding ?

Def.: Infant is given other feeding rather than breast milk in the period he naturally receive human milk.

Types: - Fresh animal milk - dried animal milk - Condensed & evaporated animal milk.

Choice depends on: - Financial conditions. - Convenience. - Location.

Indication:

- * C.I of breast Feeding
- * Complementary & supplementary feeding.
- * Mother → absent or Died .
- * Baby → twins - dis - allergy - fat intolerance – metabolic disorder.

Fresh animal milk

1- Amount :

*** Age method (rule of ten)** - 1st day, 10ml/fed/d.

- 1st week, $\uparrow$ 10ml/fed/d.

- 1st month, $\uparrow$ 10ml/fed/w

- 1st year, $\uparrow$ 10 ml/fed/m.

*** Wt method:** - Infant milk = 150 ml / kg / d.

- In emergency: B.Wt. X 20 + 20 = ... ml / fed.

*** Caloric method** - 110 cal /kg /day. - 100 ml/milk $\rightarrow$ 67 cal.

2- Modification = formula = Concentration

Aim : Approximate to Human milk.

Cured $\rightarrow$ small easy digestible.

How diluted :

1- 2 months $\rightarrow$ 1:1 (milk portion : water)

3 - 4 months $\rightarrow$ 2:1

5 - 6 months $\rightarrow$ 3:1

> 6 months $\rightarrow$ whole milk given

Sugar : flat teaspoonful 120 ml mix.

3- Frequency : as before

4- types:

Raw :

Disadvantage: contamination – large cured

Pasteurized : heating at 143 F for 1/2 hr. rapid cooling .

Advantage: destroy bacteria (not spores)

Small cured.

Certified by lab. Exam.

Humogized: Break fat globules $\rightarrow$ Small cured.

Feeding 13 / 14

*With Best Wishes
د/ مادل & د/ الجيزي*

Evaporated & condensed

Not used in EGYPT

Dried milk

Def: milk(H₂O removed +dry solids + powder form) in sterile container.

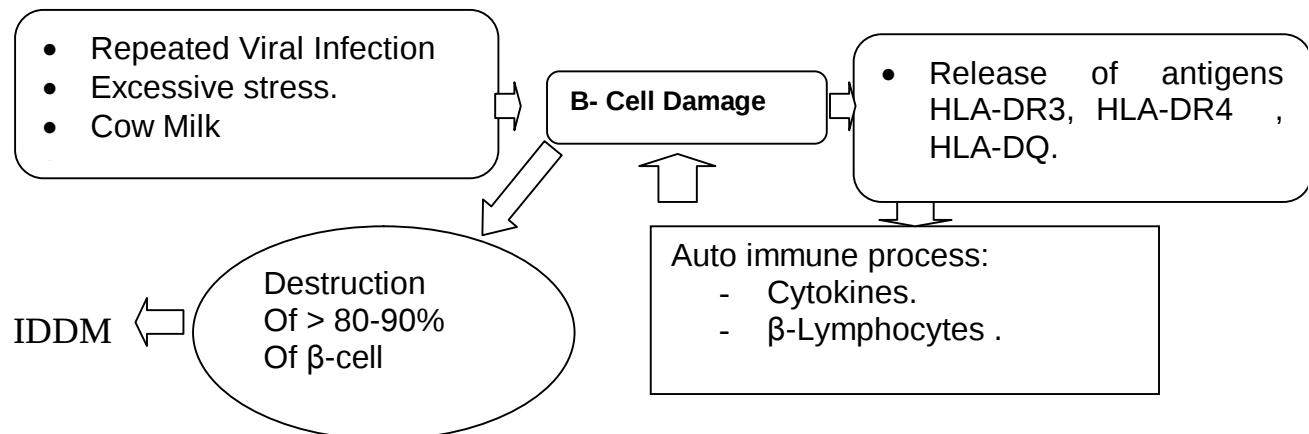
<i>Advantages</i>	<i>Preparation</i>	<i>Types</i>
- Sterile - Easy digestible - Constant composition. - Fine cured - Not require refrigerator -Fortified with Vit.	- Dil: 1:8 (milk : water) How: powder measure (4gm) + 30 cc previously boiled H₂O - Exception of dilution: premature 1st 2 week	1- Full cream (Nido). 2- 1/2 cream (Bebelac Z12) 3- Humanized (Similac). * Fat, CHO, Ptn → approximat. human. Milk. * More Unsat. F.A * Minerals → ↓ *Alter. amount of caseinogen to lactalbumin 4- Skimmed * Indication: -Fat intolerance. -PEM -Diarrheal dise. -Prematurity * Types: -Non fat skimmed (0.5% fat) -1/2 fat skimmed (1.5% fat). 5- Special types Table below

Acidified milk	Butter milk	Protein milk	Specific
Indication: - PEM - diarrheal dise - prematurity Types: 1/2 cream Full cream-skimmed Adv.: - acid →disinfectant. - cured →small	↓ fat ↓ calories	→ Disadvantage: -Expensive -Bad taste -Constipation -High protein Content -Low Caloric value	-Hypo allergic: allergy to human and animal milk -Salt free: H.F. -Lactose free Formula (Isomil) : lactose intolerance -Phenyl alanine free: (Lofenlac) Phenyl ketonuria

IDDM

1- **Def:** Chronic hyperglycemia d.to insulin deficiency or resistance to its action.

2- **Pathogenesis (immunogenetics):**



3- **C/P:**

a- **Symptoms of hyperglycemia:**

- Polyuria
- Polydipsia
- Polyphagia
- Nocturnal Enuresis.
- Monilial Infection.

b- **30% of cases presented for 1st time by DKA :** **NADH TV P**

- Neuro: . drowsiness . coma
- Abdominal pain.
- Hyperventilation
- Dehydration.
- Tachycardia
- Vomiting
- Polyuria

4- **Investigation (Monitoring):**

1- Urine analysis: kgm

- Ketone bodies.
- Glucose.
- Micro-albuminuria (D. Nephropathy).

2- Blood:

- Glucose:
 - Random (> 200 mg /dl with symptoms)
 - Fasting.
 - Post-prandial.
- Glucose tolerance test: **Give 75 gm glucose**

	Fasting mg /dl	Post-prandial
IDDM	> 110	>180
Impaired glucose intolerance .	<110	110-180

- HB A1c (3 months)
- Serum Insulin, Creatinine.
- Investigation for complications : RFT , Fundus ex.

6- Complications (Problems) of IDDM:

- a- Acute: DKA, HHNK.
- b- Chronic: Micro, Macro-angiopathy.....
- c- Complication of Insulin.

5- TTT: **D I E + F**

- Diet.
- Insulin + Future therapy.
- Exercise.

Diet :

Aim :

- 1- ↓ Symptoms.
- 2- ↓ Acute Compl. (Hyperglycemia).
- 3- ↓ Chronic Compl.
- 4- Optimal growth & development.
- 5- Optimal lipid Levels.

Dr. Madeh Dr. AlgizY

Advice:

- Avoid: drinks with sugar (drink water).
Fried, very sugary food.
- Meal: Main Part: vegetables, bread, pasta , Potatoe.
Small part: meat, egg , cheese.
- Alternative: Fish – Pulses.

Difficulties:

- 1- Ptn : Consider diet a traumatic aspect in ttt.
- 2- Doctor: Programs based on scientific not practical Basis.
- 3- Short term Penalties not apparent.

Exercise*** 1. Running 2. Cycling:**

- * At H.R 70 % of Maximum H.R (220-age in yrs)
For 30 min / 3 times / week.

*** Benefits:**

- Physiology.
- Biochemistry.
- Psychology (self –esteem).
- Improve lipid profile
- Improve sensitivity to insulin.

*** Precautions (Avoid Hypoglycemia):**

- 1- Before exercise: CHO Snack if blood glucose < 100 mg/dl.
- 2- Prolonged: 15-30 gm CHO every 30 min. during exercise.
- 3- Avoid exercise at time of peak insulin action
- 4- Ms. Injected with short acting insulin.

Insulin (Q 2006)

- 1- Types:** **A-** Beef insulin. **B-** Purified pork insulin

C- Human insulin : ☺ Recombinant DNA technology.

☺ Chemical modification of pork.

2- Advantages of Human insulin:

- Identical to human insulin.
- Less immunogenic than animal insulin.
- No insulin resistance, allergy.
- No lipodystrophy.
- Free from animal contaminants.
- Better for ptn complications.

3- Preparations:

- Short acting → Regular insulin.
- Interm. Acting → Isophane (NPH) – Lente insulin.
- Long acting → lantus.
- Mixures → Mixtard – Novomix.

4- New preparations = Insulin analogues:

- Ultra – Short acting → Insulin Lispro :
(Act after 1/4 hrs – for 4-5 hrs).
- Long acting → glargine (basal level of insulin).

5- Aim (goals):

- ↓ Symptoms.
- ↓ Acute complications (comas).
- ↓ Chronic complications .
- Improve exercise, work performance.
- Restore body mass.
- Improve sense of well being.
- ↓ Malformations of foetus.
- ↓ Infection.

6- Route:

1. S.C.
2. I.V. (regular).
3. Insulin Pump.
4. Inhalation (new).
5. Oral (new).

7- Site of injection:

1. Lat aspect of arm .
2. Lat. aspect of thigh.
3. Lower part of abdomen.

8- Factors affecting absorption:

1. Anatomical area.
2. Blood flow.
3. Size of dose.
4. Lipodystrophy.
5. Exercise.

9- Dose:

a- In non –ketotic patient:

- Start regular insulin every 4-6 hrs by sliding scale regimen.

Or

- Give 0.2-0.4 Unit/kg /day Mixtard. (2/3 dose at morning, 1/3 at night).

b- In DKA:

- Give IVI insulin 0.1 Unit/kg/hour.

- Recommended rate of blood glucose drop (50:80 mg/dl/hr).

- If $> 80 \rightarrow$ insulin dose to 0.05 U/kg/hr.

- If $< 50 \rightarrow$ insulin dose to 0.15 u/kg/hr.

- If bl. Glucose < 150 mg/dl with presence of acidosis give glucose 25% +

glucose 5% in saline 45%.

- Don't stop insulin or ↓ below 0.05 as it is essential for ketone metabolism.
- After disappearance of acidosis (12-24 hr) give S.C. insulin acc. to sliding scale (1/2 - 1 hr) before stop insulin infusion.

<i>Bl. Glucose level</i>	<i>Insulin dose</i>
< 70 mg / dl	No insulin
70-110	0.1 Unit/kg
110-140	0.2 Unit/kg
140-180	0.3 Unit/kg
180-210	0.4 Unit/kg
> 210 mg/dl	0.5 Unit/kg

10 - Side effects:

- * Allergy * Hypoglycemia * Lipodystrophy * Sick day Problem
- * Resistance * Cerebral edema * pre- operative management.
- * early morning hyperglycemia (Somogi phenomenon)

Diabetic Ketoacidosis

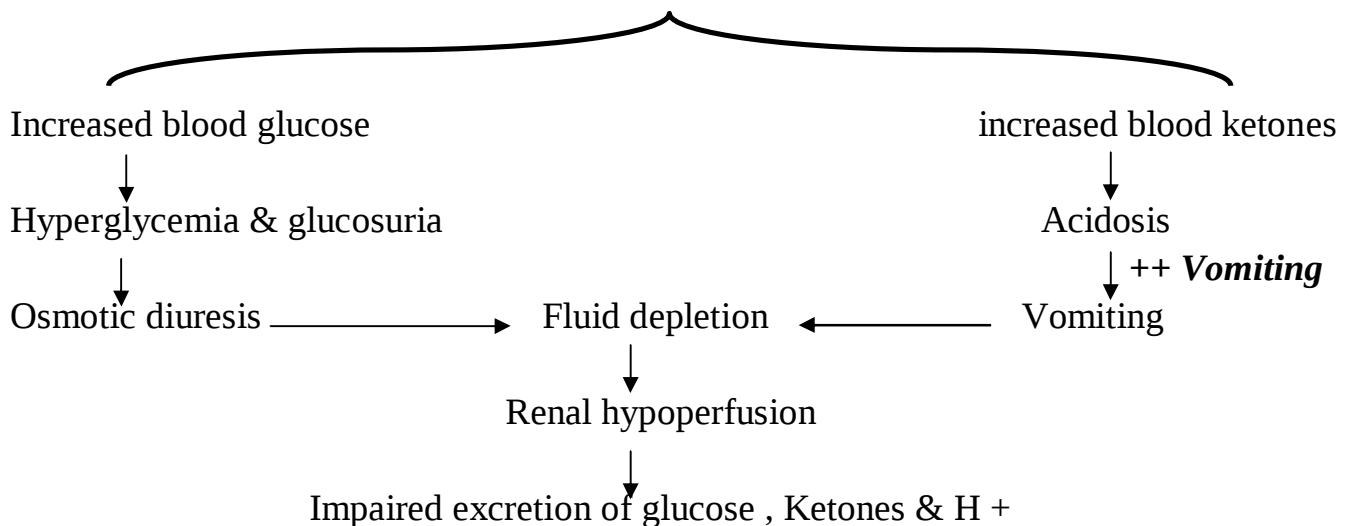
Def : - It is an acute metabolic derangement in IDDM. Characterized by hyperglycemia, acidosis & ketouria.

C/P :

- 2 فوق Vomiting
- تحت Polyuria
- 2 # Tachycardia
- Hyperventilation
- 2 Neuro Drowsiness
- Coma
- 2 مع بعض Dehydration
- Abdominal pain

Pathogenesis :

Insulin deficiency

**Diagnosis**

- Bl. Sugar > 250 mg/dl.
- Bl. PH < 7.2.
- $\text{HCO}_3^- < 15 \text{ meq./dl.}$
- Glucosuria & Ketonuria.

- It is important to note that degree of hyperglycemia does not correlate with the degree of acidosis.
- It is important to determine:
 - $\text{S. osmolality [effective]} = 2 (\text{Na} + \text{K}) + \text{Bl. Glucose .mg dl.} / 18.$
 - $\text{Corrected S.Na} = \text{Na} + 1.6 (\text{bl. Glucose} - 100) / 100$

Problems in DKA

- 1- Dehydration
- 2- Electrolyte dist
- 3- Hyperglycemia.
- 4- Acidosis.
- 5- PPT (infection)

Protocol of therapy of DKA Q أكثر سؤال بيـ بيـ**I. Confirm diagnosis:**

- a- Characteristic history & examination . C/P 2
- b- Biochemical confirmation: (Invest)
 - Glucosuria, Ketonuria.
 - á bi. Glucose.
 - S.Na, SK.

- Corrected Na = $\text{Na} + 1.6 \left[\text{bl. glucose} - 100 \right] / 100$.
- Serum osmolality = $2 [\text{Na} + \text{K}] + \text{Bl. Glucose} / 18$.
- Blood PH & HCo₃.

II. Resuscitation

In shock with poor peripheral pulse or coma.

- 1- O₂ 100 % by face mask.
- 2- Naso-gastric tube to drain stomach if there is vomiting or impaired consciousness.
- 3- Urine catheter.
- 4- Start IV saline 10 ml/ kg, given over 15 mins. (Emergency dose)

III. Rehydration:

- Start by normal saline (0.9) 20 ml / kg / H for 1 -2 hours.
- Then give fluids by the following regimen:

1- Fluids volume:

- 6 ml / kg / hr for Child Wt 3 - 9 kg,
- 5 ml / kg / hr for Child Wt 10 - 19 kg.
- 4 ml / kg / hr for Child Wt > 20 kg [max. 250 ml / hr]

2- Type of fluid:

- Depends on corrected Na level & Bl. Glucose.
 - If hyponatraemic → use saline 0.9.
 - " normo " saline 0.45 %
 - " hyper " saline 0.3 %
- Add glucose 5 % when Bl. Glucose < 300 mg / dl.
- If Bl. Glucose < 150 mg / dl & still acidosis ; add 2 ml / kg / hr glucose 25%
- Add 20 mEq K / Lit, fluid taken.

3- Warning signs & symptoms of brain edema.

- Headache.
- Bradycardia.
- Restlessness & ↓ drowsiness.
- Incontinence.
- Cranial nerve palsy.
- ↓ Bl. Pressure & ↓ O₂ saturation.

Action:

- Give Mannitol 1 gm / kg over 20 mm. & repeated every 4 hours if needed.
- Nurse child with head elevated.
- Halve rehydration infusion rate.
- Be ready for assisted ventilation.

4- Oral fluid:

- **In severe dehydration & acidosis:** only allow sips of cold water.
- **Oral fluids** :as rehydration solution, juice should be used after clinical improvement & no vomiting & subtracted from IV fluid.
- **In mild case of DKA without vomiting or sever dehydration :**
Oral rehydrationsolution can be used & volume calculated as usual.

IV. Insulin

- Not started except after shock has been Success fully reversed by emergency resuscitation
- Given by IV infusion 0.1 u/ kg / H. low dose regimen
(50 U insulin in 500cc saline : 0.1 U/q ml)
- Recommended rate of blood glucose drop : 50 - 80 mg. dl / H.
§ IF > 80 mg /dl /H: \$ dose of insulin to 0.05 U / kg / H.
§ IF < 50 mg /dl/H: #dose of insulin to 0.15 U / kg / H.
- If bl. Glucose \$ than 150 mg / dl in presence of ketoacidosis use glucose 5 % in saline 0.45 plus glucose 25 % [2 ml / kg / H]
- don't stop insulin infusion or \$ below 0.05 U / kg / H as it is essential for ketone bodies metabolism.
- After 12 - 24 hours of disappearance of acetone : start SC insulin according to sliding scale 1/2: 1 hour before stop of insulin infusion.

Sliding scale for regular insulin Dosage.

<i>Blood glucose level</i>	<i>Insulin dosage</i>
Less than 70 mg / dl	No insulin
70 – 110 mg / dl	0.1 U / K max . 4 units
110- 140 mg / dl	0.2U / kg max. 6 units
140 - 180 mg / dl	0.3 U / kg max. 8 units
180 -210 mg / dl	0.4U / kg max. 10 units
More than 210 mg / dl	0.5U / kg max. 12 units

V. Na Hco3:

- Recently has no role in treatment of DKA except in sever acidosis associated with myocardial depression.
- Dose (12- observed Hco3) x 0.3 x body wt.

VI. Treatment of precipitating factor.

Prevention of DKA

- 1- Monitoring of blood glucose & urine ketone.
- 2- Check for urine ketone if
 - bl. Glucose > 250 mg / dl.
 - child feels ill.
- 3- When ketone becomes positive mild, extra regular insulin is given until ketone disappear
- 4- Proper sick day management.

Common Errors in diagnosis & Management of DKA

- 1- **Delay** diagnosis ttt of cerebral Edema .
Delay starting insulin
- 2- **Rapid** \$of glucose
Rapid & excessive fluids.
- 3- **Stop** insulin (before S.C insulin – still acidosis)
- 4- **Urine output** as indicator of dehydration

Cretinism

A/E :

1- 1ry : Congenital :

- Agenesis, Hypoplasia, ectopia
- Dyshormonogenesis.
- Mother Taking anti – thyroid drugs ,
Or exposed to Radio- active Iodine.

Acquired:

- Hashimoto disease, surgical excision, irradiation, iodine deficiency.

2- 2ry: ↓ TSH from pituitary.

3-3ry: ↓ TRF from hypothalamus.

C/P :

*** at birth & early neonatal :**

- 1- Feeding difficulty, choking & anorexia
- 2- Constipation, abd. distention, umbilical hernia, delayed passage of meconium
- 3- Heavy birth wt. (Over wt).
- 4- Hypothermia, cold skin.
- 5- Open posterior fontanel.
- 6- Less activity, always sleep, little cry hoarse voice.
- 7- Prolonged physiological jaundice.
- 8- Bradycardia: ↓ HR (Slow Pulse) .
- 9- Apneic attacks: ↓ Respiratory rate.
- 10- X- Ray knee: absent ossific centers at birth of the lower end of the femur.

APGAR score of early suspicion of hypothyroidism

H → Hypotonia → 1

hypothyroidism مختصره في كلمه

Y → Yellow (icterus >3) → 1

P → Pallor, cold, hypothermia → 1

O → Open post. fontanel → 1

T → Tongue enlarged → 1

H → Umbilical hernia → 2

Y → absent Y (female) → 1

R → Rough dry skin → 1

O → Edematous typical face → 2

I . D. → Inactive defecation → 2

Birth wt > 3.5 kg → 1

Post.mature > 40w → 1

Total = 15.



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If score > 5 suggest hypothyroidism, must investigate.

Typical Symptoms & Signs:

A- Delayed growth & development + M.R.

- Delayed motor mile stones.
- Delayed social development.
- Growth retardation & short stature .

B- Characteristic features:

1- Head:

- o Face → coarse puffy face.
- o Skull → delayed closure of fontanel (anterior).
- o Hair → coarse dry hair, low hair line.
- o Eyes → hypertelorism, puffy eye lids, scanty hair of brows.
- o Nose → depressed nasal bridge.
- o Tongue → macroglossia, thick lips.
- o Teeth → delayed eruption, tendency to decay.

2- Neck: short & webbed. & thyroid may palpable .

3- Skin: Pale yellow skin (carotenemia). & Dry, rough, clod.

4- Abdomen: → Pott's belly abdomen. & Umbilical hernia.

5- C.V.S: **Bradycardia.**

- o **Haemic** murmur.
- o **Cardiomegally** → CHF.

6- C.N.S: Hypotonia, Hyporeflexia, apathy.

<i>Investigations</i>	<i>TTT start early to prevent M.R</i>
1- Hormonal assay	L - thyroxin : 5ug/kg/day
- T3 Level	→ ↑ the dose acc. to response up to 10 ug/kg/day
- T4 Level	till improvement or appearance of signs of

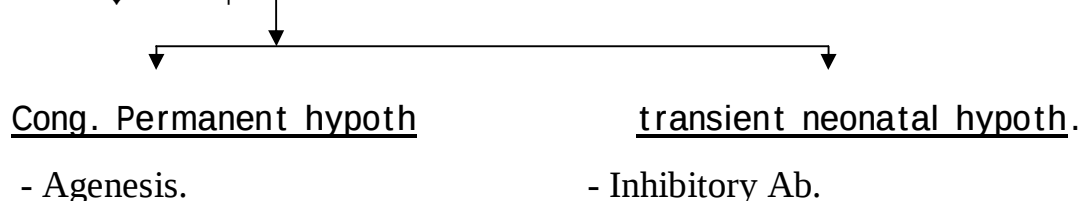
- TSH (<5Mu/ml normally). - Radioactive T3 resin uptake. - Free T4 index. - Free T3, T4, (by RIA)	hyperthyroidism. → In older age → ↓ the dose Manifestations of Overdose. - Fever. - Diarrhea - Tachycardia - Tremors - Loss of wt
2- Thyroid antibodies.	
3- Radioactive isotope studies.	
4- Thyroid U/S.	
5- National Screening.	
6- X-ray knee.	

National screening of hypothyroidism

- 1- **Value**: early diagnosis of cong. Hypothyroidism to prevent M.R.
- 2- **Method**: blood sample on filter paper collected & examined for TSH or T4.
- 3- **Time**: 3-7 days after birth to avoid TSH surge during 1st 3 days.
- 4- **Screening by TSH**: < 6.6 IU/dl → diagnostic.
- 5- **Screening by T4**:
 - > 40 IU/Ml → Hypothyroidism.
 - 20-40 IU/Ml → Suspicious.
 - < 20 IU/Ml → Normal
 - If screening +ve → re-investigate the baby for thyroid profile: total T4, free T4, TSH , Thyroid U/S.
 - Start TTT at once before results of investigation.
 - If +ve → Continue TTT.
 - If -ve → Stop TTT.

6- **Interpretation of results**:

a- ↓T4 & ↑TSH:



- Dyshormonogenesis
- Ectopia
- Iatrogenic: Anti-thyroid drugs taken by mother
- Iodine deficiency.

b- ↓T4 & normal TSH:

- ↓ TBG (thyroxin binding globulin).
- Error in sampling.
- Euthyroid sex syndrome.

c- Normal T4 & ↑ TSH:

- Compensated transient hypothyroidism.
- Drugs & dyshormonogenesis.
- Ectopia.

Ca hemostasis

Normal serum Ca → 8.5 -10.5 mg / dl.

شفوى هام جدا د / زكريا الزينى

Normal ionized Ca mg/dl 4.5-5 mg/dl.

Hormonal control of Ca level:

1- Parathyroid hormone:

- ↑ Ca absorption from the intestine by stimulation of calbentine protein.
- ↑ Ca reabsorption & P excretion in the renal tubules.
- ↑ Activation of Vit D in the kidney to 1.25 DHCC.

2- Vit D:

- ↑ Ca & P absorption from the intestine.
- Essential for mineralization & resorption of the bone.

3- Calcitonin: ↓ Ca level by ↑ Ca deposition in the bone & ↓ bone resorption.

TETANY

Def: a state of hyperexcitability of myoneural junction due to ↓ ionized Ca in the blood.

A/E:

1. ↓ PTH 2. ↓ Vit D. 3. ↓ Mg. 4. ↑ Inorganic P.

1. ↓ Parathyroid hormone.

- A. Transient neonatal hypoparathyroidism: due to maternal hyperparathyroidism & early cow milk.
- B. Congenital aplasia or hypoplasia.
- C. Familial hypoparathyroidism either X linked or AD.
- D. Autoimmune hypoparathyroidism.
- F. Surgical removal as in Grave's dse.
- G. Pseudo-hypoparathyroidism: unresponsiveness to PTH.

2. ↓ Vit D:

- A. Infantile rickets.
- B. Malabsorption due to celiac dse & chronic parasitic infestation.
- C. Impaired synthesis of active Vit D due to renal rickets.

3. ↓ Mg:

- A. ↓ parathyroid hormone secretion as in familial hypomagnesemia.
- B. Malabsorption.

4- ↑ Inorganic P:

- A. Organophosphorous compound poisoning.
- B. During TTT of leukemia.

C/P:

* **Symptoms** : muscle pain, cramps, stiffness, numbness Tingling in the hands & feet.

* **Signs:**

A. Latent tetany : if the total serum Ca 7 : 9 mg/dl. , the ionized Ca 3: 4 mg/dl.

- 1. Chvostek`s sign: tapping of the facial nerve in front of the ear → spasm of the facial ms in the same side.
- 2. Trousseau's sign: compressing the upper arm with a sphingmanometer cuff with

pressure midway bet. Systolic & diastolic BP → carpal spasm of the hand.

3. Erb`s sign: ms. contraction can be initiated by galvanic current less than 6 milliamperes.

4. Peroneal sign: tapping on the peroneal nerve → pedal spasm .

B. Manifest tetany:

If total Ca < 7.5 mg/dl or ionized Ca <3.5 mg/dl.

1- Carpal spasm: Flexion of the rest & metacarpophalangeal joints.

Extension of the interphalangeal joints.

The thumb is adducted & draw into the cupped palm.

2- Pedal spasm: Extension of the ankle with planter flexion of the toes the patient fall on his face.

3- Laryngeal spasm: attacks of spasmodic closure of the glottis → stridor.

4- Convulsions: generalized tonic or tonic – clonic with loss of consciousness In advanced stage.

Investigations:

1- S. Ca total & ionized.

2- S.Albumin & pH.

3- Mg & P.

4- Alk.P.

5- X. ray of long bone.

6- Renal function tests e.g. Creatinine clearance.

7- PTH estimation.

Treatment:

1- **Ca gluconate** 10% (90 mg elemental Ca/ml).

0.5: 1ml / kg IV at a rate of 0.5: 1ml/min.

Repeated when needed & may be maintained at 1 ml/hr.

2- **Vit D:** D2: 0.1: 0.5 mg/d. D3: 0.01: 0.1 mg/kg/d.

1.25 DHCC: 0.25 ug/d.

Short stature

def: Height of growing child below 3rd percentile for age.

Factors affecting height & growth:

- age - sex - Race
- nutrition
- chronic Dse
- chronic endocrinal dise.
- Hereditary (Mid Parental Height =

$$[\text{Mother} + \text{father} (+ 13 \text{ if male \& -13 if female})] / 2 (\pm 5)$$

Causes:

A- Notrnal growth variants:

<i>Familial</i>	<i>Constitutional</i>
* Child grow within expected MPH - 5 * Bone age = Chronological age.	* Period of delayed growth in 2 nd year . * + ve family Hx of delayed puberty. * TTT: testosterone.

b- Syndromes:

- Down
- Noonan.
- Progeria.
- Turner
- Sickle.

c- Chronic diseases & TTT:

- Chronic renal dise.
- Malignancy
- Chronic inflame. bowel dise.
- Rickets.
- Chronic Pulm. dise (cystic fibrosis).
- Skeletal dysplasia
- Chr. Corticosteroid ttt
- (osteogenesis imperfecta).

d- Endocrinal disease:

- Hypothyroidism.
- G. H. ↓↓.

- Pseudo hypoparathyroidism. - ↑↑ Glucocorticoids.

Pituitary dwarfism

A/E.:

- Congenital: agenesis, aplasia, dysplasia.
- Traumatic.
- Infection: T.B., sarcoidosis.
- Neoplastic: cranio-pharyngioma, pituitary adenoma.
- Iatrogenic: surgical excision, irradiation.

C/P.:

a- At birth: * normal size & wt.

* may be hypotonia, hypoglycemia.

b- During childhood:

- * Short stature.
- * Rounded head.
- * Prominent head.
- * Broad face.
- * Depressed nasal bridge.
- * Saddle small nose.
- * Undeveloped Mandible & chin.
- * Short neck.
- * Proportionate limbs with small hand & feet.
- * Underdeveloped genitalia & absent body hair.
- * Normal IQ.

c- Cases with destructive pituitary lesion:

- * Initially normal, then growth failure.
- * Multiple hormonal ↓: - TSH – ACTH – ADH – GnTH - GH

* Headache, vomiting, squint.

Investigation.: as known:

+ appropriate measurement to diagnose short stature. + IGF1. & GHRF.

TTT: - Of the cause.

- Recombinat human growth hormone (r HGH). S.C
- Replacement therapy for other hormones.

Causes of delayed puberty???

a- Hypogonadotropic conditions	b- Hypergonadotropic conditions
1- Multiple pituitary horm. ↓. 2- Isolated G.H. ↓. 3- Isolated GnTH ↓ 4- Syndromes (Pader - Willi syndr). 5- Systemic dise.: Nutritional, psychologic. 6- Other endocrinal cause: hypothyroidism. Hyperprolactinemia. 7- Polycystic ovary dise. 8- Constitutional delay in growth.	1- Ovarian and testicular dysgenesis. 2- Enzyme defect. 3- Androgen in sensitivity. 4- Gonadal toxins or radiation ttt. 5- Other miscellaneous causes.

Casues of precocious puberty????

Central:

- Idiopathic true precoious pouberty.
- CNS disorders (meningitis, encephalitis) .
- CNS tumors (Hamartoma, hypothalam. Tumor).

Peripheral:

Male	Female
A- Adrenal tumors	* Adrenal tumors.
C- Congenital adrenal hyperplasia.	* Ovarian tumors.

L- Leyding cell testicular tumor.	* Follicular cysts.
T- Testotoxicosis.	* Exogenous estrogen.

ACUTE VIRAL HEPATITIS

AE : * Hepatotropic virus. : A , B , C , D , E

* Non H.V. : - Adenovirus - EBV CMV. - Coxaki V.

	A	B	C	E
Type	RNA	DNA	RNA	
Mode of Trans.	- Feco – oral - Contaminated Water , food	1. prenatal transmtion: Factors aff. Develop of HBs Ag in infant Maternal → HBs Ag Titre ↓ HBe Ag +ve 2. during Birth 3. transplacental 4. parenteral	1. perennial The most important 2. Heterosexual 3. Occupational 4. BL. Trans. 5. I.V Drug Abuse	Feco-oral
IP	1 M`	1- 6 M`	1/2 - 6 M`	1 M`
C/P	<u>Symp</u> * F A H M * GIT (A,N,V) * Pruritis * Jaundice <u>Signs</u> * Liver : - Tender - Enlarge. * Spleen : Enlarge.	<u>Prodroma</u> * F A H M , LN pathy * Serum sickness like "Arthralgia urticaria Arthritis , M.P rash " <u>Icteric Phase</u> * 2 wk after prodroma * Pruritis * liver: ↑ , Tender * Jaundice * Spleen : ↑	- Fatigue الاله - Abd. Pain - A N V M H ↓ Malaise headache	
Comp	- Fulminant hepatitis - Ch. Hepatitis : Never - Carrier : Never	- Ch. Hepatitis - H.C.C - Carrier	→ → →	→ - Never - Never - No Carri.
Inv	1. <u>HAV Igm</u> : . Acute Phase. . Disappear after 4 M 2. <u>HAV IgG</u> : . convalescence Phase . . presist for years . 3. stool exam → Virus 4. SGOT, SGPT : ↑30 time	<u>HB Markers</u> . HBs Ag, HBs Ab . HBe Ab, HBc Ab . HBe Ag . HBV DNA . Anti HBC Igm , IgG	1. ELIZA : 1,2,3 2. PCR : HCV RNA 3. ↑ SGOT, SGPT	1. <u>HEV Igm</u> Recent infec. 2. <u>HEV IgG</u> Past inf 3. <u>School exam</u> : Detect Virus
Prognosis	<u>Chronosity</u> : Never <u>Carrier</u> : Never <u>Cancer</u> : Never	+ + +	++ ++ ++	Never Never Never

FULMINANT HEPATITIS

Complication يكتب مع ال

incidence

It may complicate acute viral hepatitis

HAV HBV HCV HEV HDV most common HEV

Onset

Within 8 weeks of appearance of first symptoms of viral hepatitis & in absence of preexisting liver disease.

C/P

1. Mental changes and personality irritability
2. Deep Jaundice
3. Anorexia, nausea , vomiting
4. Fever
5. Shrunk liver & ascites.

INV

1. liver enzymes
2. ↑ PT unresponsive to vit k administration.
3. ↑ Blood ammonia.

TTT

1. Hospitalization
2. Supportive TTT till liver transplantation occurs.

Q : ACUTE VIRAL HEPATITIS MANAGEMENT

No specific treatment, only supportive treatment.

A . Goals of management

1. **Early detection** of acute viral hepatitis (C/P , INV.) يذكروا
2. **Monitoring** during acute stage to discover as early as possible fulminant hepatitis ?
3. **Prevention** of the spread of the disease
 - Careful screening & selection of blood donors .
 - TTT of chronic infection (carriers).
 - Immunization.
4. **Recognition** of The development of fulminant or chronic liver Dse.

B . General lines

1. **Bed rest** : may be only needed during early symptomatic stage.
2. **Diet**: No specific diet but you can give diet rich in vit C & give Vit k.
3. **Hospitalization**: fulminant hepatitis , coagulopathy, persistent vomiting , bleeding .
4. **Return to school** : as soon as feeling well.
5. **Follow up studies** : to ensure clearance of virus & biochemical Resolution

C . Prophylaxis :

	Passive immunization	Active immunization
HAV	ISG (Standard serum globulins) Indicated in acute HAV infect & House hold contact Dose : 0.02 ml / kg	HAV vaccine which is formalin killed vaccine 3 doses 0.5 ml IM (0 , 1 , 6 M`)
HBV	* HBIG (Hepatitis B immunoglobulin) - Indications : 1. Post exposure : Sexual & house hold contact 2. perinatal exposure - Dose : 0.02 ml / kg - Protection : for 3- 6 M`	* HBV Vaccine ; Types : 1. Plasma derived vaccine (inactivated HBs Ag obtained from plasma of chronic HBV patient) . 2. Recombinant DNA vaccine . * Dose : 3 IM doses (0 , 1 , 5 m`) * Protection at least 5 years.
HCV & HEV	Not available	Not available

	Pathology : - Steatosis - Lobular inflammation, - Cirrhosis
	Prognosis : 50 % of Acute → chronic. 20% of Ch → cirrhosis
Pathogenesis : immune mechanism → T cells Response	
TTT: goals : - ↓ viral load . - Amelioratn of hepatic dysfunctn 1- interferon alpha : • Dose : same • Markers for Response * -ve → HBV DNA by PCR → HBE Ag → HBs Ag * S. ALT → Normal • Mechanism ↓ Antiviral ↓ Anti fibrinogenic ↓ V. replication ↓ V. assembly ↓ immunologic ↑ Natural killer activity ↑ Maturation of cytotoxic T cells • S.E - Influenza like illness , fatigue , fever, chills, headache subside in 2-3 wk. - Alopecia, Depression. - Leucopenia – Thrombocytopenia • C.I - Major psychiatric lesion - Th.c < 50.000 / ccm3 - LCF • Monitoring therapy by ALT, AST, CBC 2- Luemeudine : 15 mg / kg / d orally.	TTT 1- interferon alpha • Dose : 3mulm 3 / wk For 6M S.C • Markers for Response - S.ALT → normal - S.HCV RNA → -Ve by PCR REs. Rate 40 % → response 50 % of Res → Relapse 2- Interferon alpha + Ribaverin

Q : CLINICAL EVALUATION OF CHL HEPATITIS ?

Def

AE

1 - Is it Hepatitis or not ?

• Vague symptoms : in risk group :

- 1 . A NV F M
- 2 . Abd. Pain
- 3 . Fatigability

- 1 . Dialysis
- 2 . Drug
- 3 . Bl. Transfusion

- 4 . +ve f. M
- 5 . BM Aspiration

- Hepatic Symptoms: - Ascites , Odema , Bleeding , Jaundice
- Extrahepatic symptoms: - vasculitis , PAN , Rash , Nephrotic , syndrome

2 - Is it Acute or chronic ?

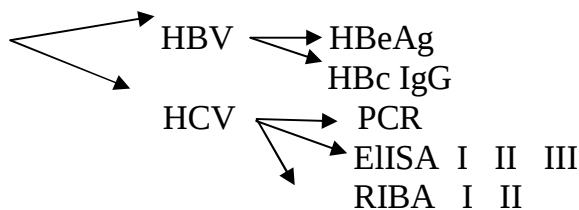
= C/P suggesting chronicity : * History & Examination & Inv

3 - What is the AE ? = Inv. Of child e` chronic Liver Dse.

1- Initial : CBC – RGT , ALT , PT , S. albumin ^{الاهم}

2- Specific :

- V. Markers



- Autoimmune study ANCA – ALKA – AMA – ASMA
- Metabolic study
 - * alpha 1 Antitrypsin level → alpha 1 AT deficiency
 - * S. Ceruloplasmine → Wilson's Dse
 - * Liver Biopsy (Glycogen storage Dse & Gaucher Dse)
 - * Urine reducing substance → Galactosemia
 - * Sweat CL → cystic fibrosis
- U/S (Abd – liver – portal vein) .
- Upper GIT Endoscopy → for bleeding.
- Liver Biopsy : * Chronic Persistent hepatitis :
 1. Intact limiting plate
 2. Normal architecture.
- * Chronic active hepatitis :
 1. piece meal Necrosis
 2. lost architecture.

AutoImmune Hepatitis

Metabolic Hepatitis

Def: progressive inflammatory Disorders.

- +ve (IgG & Auto Ab)
- exclusion of (3) :
 - Wilson Dse.
 - HCV
 - Drug induced Hepatitis

Classification : Depend on Auto . Ab.

Type 1

- * +ve ANA
- * +ve ASMA

Type 2

- * +ve Anti
- * +ve LKM

D : - Biopsy : Ø
 - Ø criteria: C/P & Exclusion of (3)

Presentatn

- Hepatitis
- * Acute (prolonged)
 - * Ch (compensated)
 - * Fluminant (encephalopathy)
 - * LCF (no encephalopathy)

Management

Indicatr

- * Absolute - S.AST > 10 time
- Necrosis
- * Relative - S.AST, less. < Absolute
- Jaundice, fatigue, arthralgia

Stop ttt criteria :

- -ve IgG → 1 y`
- ↓ Auto Ab → Check 3 / m`
- Biopsy → No inflammation

TTT

Prednisone : 1mg / kg / d single Dose
 Azathioprine : 1mg / kg / d single Dose

Monitoring : ALT, AST , CBC

1. α1 Antitrypsine deficiency :

- Ø - α1 Antitrypsine phenotyping.
 - Biopsy : α1 An.t. Globules +ve PAS
 ttt: Neonatal cholestasis.
 P.H.
 Liver Transplantation.

2. Gaucher`s Dse:

Glucocerebrosidas level in WBCs

- Liver Biopsy → Foam Cells , fibrosis.
- Anemia
- Thrombocytpnia.
- HSM
- Genotyping (L444p , L444p)

D : * enz .replacement therapy
 * Wilson`s Dse.

ttt: 1. Enz. Replacement Therapy :

Goal: HB : Normal
 Platlets: Normal
 Prevent splenomegally
 Prevent bone crisis

2. B.M Transplantation .

3. Wilson Dse :

C/P: * sever hepatic dysfunction .

- * neuro : - After 12 y` old
- Kayser Flisher ring
- Artheritis
- Acute hemolytic crises

Lab : decrease S. ceruloplasmine ,
 increase cupper (liver urine)

ttt : - penicillamine 500 mg / d
 - TETA 500 mg / d

4 -Galactosenia :

C/P: - Cataract

- Urine : +ve glucose oxidase tes

ttt: Glucose free diet .

5- Glucogen storage Dse:

ttt : Nocturnal NG Infusion

Cirrhosis

Def : Diffuse liver process characterized by Fibrosis – loss of Architecture- Abn. Nodules
A / E :

<u>1- Infections</u> <u>i Viral infections</u> - Chronic HBV : 15 % cirrhosis - Chronic HCV : 25% cirrhosis - Chronic HDV : 70 – 80 % cirrhosis <u>ii Bacterial infections</u> eg. Ascending cholangitis. <u>2- Nutritional causes</u> - i PEM (protein energy malnutrition) - ii TPN (Total parenteral nutrition) - iii Hypervitaminosis <u>3- Metabolic causes</u> - i - 1 α AT deficiency - ii - Wilson's disease. - iii - Gaucher's syndrome. - iv - Galactosemia. - v - Cystic fibrosisetc. <u>4- Toxic causes :</u> Hepatotoxic drugs : - methyl dopa - Methotrexat -Chlorpromazine <u>5- Autoimmune disorders.</u> - I . inflammatory bowel diseases eg. Ulcerative colitis - ii . primary sclerosing cholangitis iii. plasma cell hepatitis	<u>6- Idiopathic causes</u> eg. familial intrahepatic cholestasis <u>7- Vascular causes</u> - i . Venous occlusive disease : obst. hepatic veins inside liver - ii . Budd-Chiari syndrome : obst. Hepatic veins entering - iii . IVC thrombosis. - iv . Pericardial - v . Constrictive pericarditis - vi . Right sided HF <u>8- Biliary malformation</u> - i - Alagille syndrome - ii- Ascending cholangitis - iii- Biliary atresia, stenosis - iv- paucity (Non Alagille) - v - choledochal cyst - vi - congenital hepatic fibrosis <u>9- others</u> - i cystic fibrosis of pancreas - ii Drugs like what ? NB 1-6 : Hepatic causes 7 : Post hepatic causes 8-9 : prehepatic causes
---	--

C / P

It depends upon i predomineni process.

The predominant process either : - Necrosis , - fibrosis and regenerating

* If necrosis > regeneration → parynchymatous decompensation. →
hepotocellular failure which is manifested by :

* Jaundice

* Bleeding Tebdency

* Abd. Pain , nausea , vomiting

* Anorexia, fatigability

* If Fibrosis > necrosis → Vascular decompensation →
portal hypertension which is manifested by :

* Hypersplenism * Bleeding varices * Ascitis & oedema Of L.Limbs.

C / P of Liver cirrhosis

1- C/P of compensated cirrhosis

I - No stigmata of chronic liver disease .

II - Hepatomegally (what are causes of enlarged cirrhotic liver)

III - Splenomegally

IV - Mild elevation of transaminases

V - Prolonged anorxia & fatigability may be i only symptoms in compensated cirrhosis.

2 - C / P of decompensated cirrhosis

A) Parynchymatous decompensation

Hepatic Manifestations J A B P

- 1- **J**aundice
- 2- **A**norexia , nausea, vomiting
- 3- **A**bdominal pain
- 4- **A**bdomianl enlargement (liver – spleen)
- 5- **A**ltered consciousness
- 6- **B**leeding tendency
- 7- **P**ruritis

Extrahepatic Manifestations

- 1 - General
- 2 - Dematological
- 3 - GIT
- 4 - Haematological
- 5 - CVS
- 6 - Endocrine
- 7- Neurological
- 8 - Nuitrional
- 9 - C/p of complications

B) Vascular decompensation = portal hypertension

- 1- Splenomegally is cardinal for diagnosis .
- 2- Collaterals between systemic & portal circulation .

- Oesophageal varices → GIT hge
 - Caput medusa
 - Hemorrhoids → bleeding per rectum
- 3- Ascitis & Oedema. Lower limbs.

BEST WISHES
د/مادح & د/الجزى

2 . C/P of decompensation cirrhosis

A) Parynchymatous decompensation

ii - Extra hepatic manifestctions

- 1) General :**
- Failure of health
 - Failure to thrive
 - Fatigability
 - Fever (continuous – low grade)

- 2) Dermatological :**
- purpura
 - Pruritis
 - Spider naevi
 - Palmar erythema
 - Paper- money skin

- 3) CIT :**
- Varices
 - Haemetemesis / melena
 - Peptic ulcer disease
 - Gastrooesoph. reflux GER
 - Steatorrhea

4) Haematological

* **Anaemia:** caused by i following :

- BL loss (Hemoptsis / melena)
- Haemolysis (Hypersplenism)
- ↓ Fe, folic acid (malabsorplion)
- Malnuitrion (d.t anorexia)

* **Coagulopathy :**

- Decrease Synthesis of liver derived clotting proteins , prothrombln VII, IV
- Thrombocytopenia (hypersplenism)
- Vit K deficiency (malabsorption)
- ↑ Consumption of clotting factors (DIC)

- 5- C.V.S :**
- High COPD
 - Low diastolic pressure
 - Water hummer pulse

6- Endocrinological :

- DM
- Delayed puberty
- Feminization & gynaecomastia
- Inappropriate secretion of ADH presented as hyponatremia

7 - Neurological : Encephalopathy**8 - Nutritional :** - Malabsorption - Malnutrition - Anorexia - steatorrhea**9 - C / p of complications**

- Hepatorenal syndrome Presented by anuria or oliguria
- Portopulmonary syndrome presented by (Dyspnea – cyanosis – clubbing)
- Pathogenesis (A.V shunts - ↓ pulm perfusion - ↓ vital capacity d.t ascitis)

Q : CLINICAL EVALUATION OF LIVER CIRRHOSIS ?

Def

A) By C/p : (Compensated - Decompensated) شرح اسماء فقط

B) By investigations :

1- is it cirrhosis or not ?

By liver biopsy must proceed by (platelet count – P.T)

2- If it is cirrhosis what is its cause?

- For
- Viral viral markers
 - Metabolic
 - Auto immune disorders : ANA – ASMA _ ALKA _ AMA
 - Biliary Malformation : U/ S
 - Hepatotoxic drugs : Assay in Bl.

3- IS it acute or chronic ?

- By ALT - High ↑ —————→ Acute
 - Moderate ↑ —————→ Chronic

4- Is it compensated or not ?*** Pseudotumorous Decompensation**

- PT : ↑
- S. Albumin : ↓
- S. bilirubin : ↑

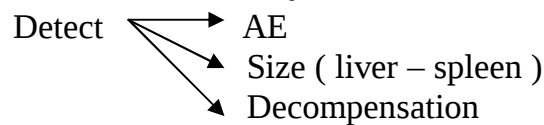
*** Vascular De. Compensation**

- portal hemodynamic .
- U/S splenomegally Ascites
- Upper GIT endoscopy varices

5- Is it complicated or not?

- U/S For → Ascites
- Endoscopy For → Varices
- Urine Na For → Hepato.Renal syndrome
- S. Ammonia For → Hepatic encephalopathy.

* *Role of U/S in cirrhosis.* شفوي



Complications of liver cirrhosis

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د / مادح & د / الجيزي

- 1- LCF
- 2- portal Hypertension
- 3- Ascites
- 4- Esophageal varices
- 5- Encephalopathy
- 6- Hepato – renal syndrome (HRS)
- 7- Spontaneous Bacterial peritonitis (SBP)
- 8- Porto- pulmonary syndrome.
- 9- GIT Hge
- 10- BM failure —→ Anemia. Growth retardation.

Management of GIT bleeding (Oesophageal varices)

1) Ass clinical status

A) clinical assessment

- * **Pulse** : Loss of intervascular volume
 - * **20 %** : rate > 120 b / min
 - * **20 – 25 %** : rate > 150 b / min
 - * **> 25%** : prolonged capillary refill
 - * **> 40 %** : no ↓ pulse, ↓ cerebral perfusion, coma
- * **Bl.p** : ↓ diastolic > 10 mm Hg on going from lying to standing when > 20 % of intervascular vol. Lost
- * **Temp** : ↓ with progressive shock
- * **R.R** : ↑ in acidosis
- * **Preph Cyanosis** : dt preph. V.C
- * **Urine output & fluid intake**: oliguria.

- 4 Vital
 - P.cyanosis
 - Urine

- B) Lab. Assessment**
- HCT, HB
 - ABG
 - bl. Loss in stool
 - Nasogastric tube to Assess bleeding Rate

c) D of bleeding site :

- | | |
|---|-------------------------|
| 1- NG tube to diff. upper , lower bleed | 2- upper GIT endoscopy. |
| 3- Barium enema | 4- colonoscopy |
| 5- Meckl's scan | 6- upper GIT radiology. |

	SBP	2ry
PMNS	> 250 cell / mm	>
PTn	< 1 g / dl	>
Glucose	> 50 mg / dt	<
LDH	serum level	> serum level
Culture	Single species	Polymicrobial

TTT

* Cefotaxime : O f C h o i c e
10 d

* Amoxillien

DD ATN – Prerenal Azotemia**A) prophylxis**

- 1- Avoid ppt – nephretic drugs
- 2- ut AE of RF

B) ttt HRS

- Volum expansion هـام
- Diet : ↓ protine هـام
- Correct electrolytes Disturbance
- Liver transplantation

Prognisis : 90 % Die.

Neonatal cholestasis

Def :

Reduction in canallcular bile flow with retention of substances which are dependent on bile flow as bile with histopathological features reflecting the nature & degree of disturbances.

Aetiology (D.D)

1- Obstructive Cholestasis * Biliary - Atrosia * 2 C - Choledocal cyst.
- Stone - Cholangitis.

2- Cholestasis with ductal paucity * Allagille's syndrome. * non syndromic.

3- Hepatocellular cholestasis * Hepatitis
* 1 Antitypsin deficiency
* Cholestasis - Drug induced - Familial.

4- Idiopathic

Comparison () idiopathic neonatal hepatitis & biliary atresia (BA)

(The most frequent causes → 75 % of cases)

	<i>Idiopathic neonatal hepatitis (hepatocellular)</i>		<i>Biliary atresia (obstructive)</i>
<i>Types</i>			a- Fetal : Jaundice since birth b- prenatal : delayed jaundice
<i>Aetiology</i>	Idiopathic		- Fetal type : cong. Anomaly - Perinatal type : infection with CMV type A.
<i>C/ P</i>	1- Jaundice	* Within the 1 st week	* At birth but may be delayed up to 3 rd – 5 th week of life.
	2- Liver	* Firm- enlarged .	Firm – enlarged
	3- spleen	* Firm – enlarged	Firm – enlarged
	4- stool	* Acholic ()	Acholic
	5- Others	* Failure to thrive * Bleeding tendency	Polysplenia . asplenia Dextrocardia
	a- Serum bilirubin	Mild to moderate ↑↑	Mild to moderate ↑↑
	b- S.bile Acids	Marked ↑↑	Marked ↑↑
	c-Alk. Phosphatase	Mild ↑↑	Mild ↑↑
	d- Trans aminase	Mild to Mod. ↑	Mild to Mod ↑

<i>Liver Biopsy (Pathology)</i>	1- Parenchyma	* loss of architecture	* intact early * preserved architecture
	2- portal tract	* Inflammatory cells infiltration * no bile duct proliferation	* Bile duct proliferation. * No or little cellular infiltration
	3- Bile stasis	* In hepatocytes & canaliculi. * Absent in portal tract	Bile plug in portal tract (specific)
<i>Radionuclide imaging</i>		- Poor intake with + ve excretion (normal passage to the intestines)	- Good intake with - ve excretion (no passage to the intestines)
<i>Prognosis</i>		* Difficulties to estimate	* 33% survival rate over 10 years.



Q : Evaluation of an infant with cholestasis
(= evaluation of an infant with acholic stool or white clay stool) نظری

- 1- Definition of cholestasis : (see before)
- 2- Aetiology (D.D) of cholestasis : (see before)
- 3- Staged evaluation of neonatal cholestasis :

a- D.D from physiologic breast milk jaundice:

- | | |
|-----------------|-----------------|
| 1- History | 5- P. T |
| 2- Examination | 6- Transaminase |
| 3- S. bilirubin | 7- Stool colour |
| 4- S. albumine | |

b. Exclusion of treatable disorders:

1. Bilateral cultures (blood & urine).
2. Viral serology (HBs Ag - VDRL)
3. α 1 antitrypsin phenotype or level.
4. Sweat chloride test: for cystic fibrosis.

C- Differentiation () extrahepatic biliary obstruction & intrahepatic disorders :

1. U/S
2. Hepatobiliary scintigraphy
3. Liver biopsy.

4- History & physical examination:

A- irritability poor feeding & vomiting :

- * Generalized infection.
- * Metabolic disorders (galactosemia - tyrosinemia)

B- Vertebral anomalies eye anomalies & murmur of peripheral pulmonic stenosis ,facial anomalies (associated anomalies) :

- * Ductal paucity. (Alagille'S syndrome)

C- Stool examination:

- * Acholic stool → if persistent → suggest BA for 10 days.
- * Pigmented stool → exclude BA.

5- General C/P:

1- regurgitation of bile content into blood

- * Bile acids → Severe pruritis & bradycardia
- * Cholesterol → Xanthomatosis
- * Bilirubin → Conjugated jaundice (olive green jaundice).

2- Accumulation of bile acids in the liver → damage

- * Portal hypertension : Ascites & Oesophageal varices.
- * Liver failure : End stage liver disease.

3- reduction of bile flow to the intestine → malabsorption

- * Malnutrition (growth retardation)
- * Diarrhea & Ca loss.
- * Loss of fat soluble vitamins :
 - Vit A : Night blindness & hyperkeratosis
 - D : Osteopenia
 - K : Bleeding tendency

6- Investigation**A) Laboratory investigations :**

- 1- Urine – reducing substances (+ ve in galactosemia)
- 2- ↓ RBC galactose -1- phosphate uridyl transferase activity = galactosemia
- 3- ↑ Serum succinylacetone, succinylacetoacetic (tyrosinemia)
- 4- Alpha – antitrypsin phenotyping for alpha -1 antitrypsin deficiency
- 5- TORCH : low diagnostic yield / culture is more specific.
- 6- Duodenal intubation with analysis of fluid for bilirubin content : green or pigmented fluid exclude BA

b) Radiological

- 1- U / S : choledochal cyst. / Absence of a gallbladder → BA
- 2- Radionuclide imaging : TC labeled iminodiacetic acid :

*** BA :**

- Radioisotope uptake by hepatocytes is good.
- Absent intestinal excretion

*** Neonatal hepatitis :**

- Radioisotope uptake by hepatocytes is delayed
- Intestinal excretion is + ve
- Pretreatment with oral Phenobarbital (5mg / kg / day) for 5 days enhances biliary excretion of the isotope and increases sensitivity 94 %

c) PTC : (Percutaneous Transhepatic Cholangiography)

d) ERCP : (Endoscopic Retrograde Cholangio Pancreatography)

e) Liver biopsy : Must be preceded by platelet count & prothrombin time
Diagnostic investigation

Management of Biliary Atresia :**1- Correctable lesions.**

- Distal atresia with patent proximal portion of the extra hepatic duct.
- Direct drainage of the biliary system into the intestine.

2- Non- correctable lesions:

- Obstruction at or above the porta hepatic.
- Direct drainage of the biliary system into the intestines (Kasai procedure.)
- Liver transplantation

Prognosis : Overallly 10 years survival rate – 33%

TTT of complications of neonatal cholestasis :

1 - Malabsorption :

Treated by : Short chain fatty acids which don't need bile.

2 - Pruritis :

A- phenobarbitone

B- Antihistaminics

C- Cholestyramin

D- Biliary diversion of bile

3 - Portal hypertension & Varices :

* Endoscopic sclerotherapy

* Endoscopic band ligation

4 - Liver cirrhosis :

* If the pt. < 2m` → Kasai procedure.

* If the pt. > 2m` → Liver transplantation

BEST WISHES

د/مادح & د/الجزی

0: COMPLICATION OF G.I OR DEHYDRATION

Met. Acidosis	↓ K	C.V.S	Renal	Post Acidotic Tetany
<u>A/E :</u> 1. HCO ₃ : - ↑ loss - ↓ reabs 2. ↑ Lactic acid 3. ↑ Keton bodies	<u>A/E :</u> 1- Loss (اسهال) 2- Iatrogenic.	<u>A/E :</u> 1. shock dt Dehydration 2. Thrombosis : dt Sever Dehydration 3. Pul. Oedema: rapid IV Infusion	<u>A/E :</u> 1. Renal Venous Thrombosis . 2. RF : dt: - ↓ renal perfusion - hypovolemia <u>TTT:</u> - IV volum expansion	<u>A/E :</u> Acidosis correction : shift Ca out bone . C/P: 1. Tetany , 2. Convulsion <u>TTT :</u> 10% Ca gluconate IV slowly
<u>D :</u> - HCO ₃ - PH - PCO ₃ - Dyspea - A N V A V - Myocardial Depression - Coma	<u>D :</u> - Hyporeflexia - Hypotonia - Arrhythmia - ECG: Inverted T		C.N.S 1. Convulsion dt * ↓ Or ↑ Na, * DIC * ↓Ca 2. Brain Damage In Na Dehydration <u>TTT :</u> - Of Cause - Anticonvulsant - proper correction of dehydration , acidosis.	
<u>TTT :</u> 1. Mild : No HCO ₃ req. 2. HCO₃ < 8 mg/dl : (HCO ₃ desired – HCO ₃ measured) X wt X 0.6 3. Sever (Resist ttt) : Dialysis .	<u>TTT :</u> - IV KCL 15% - Oral k 30 Eq/d	<u>TTT :</u> Proper correction of : - Dehydration - Shock - Slow IV Infusion	DIC C/P: - Bleeding - Anemia - gangrene - Hematemesis INV: - ↑ PT - ↑ APTT - ↓ FDP > 40 mg/dl - Throm.cyt. < 150.000/cc	GIT - Presistant Diarrhea - Post GE Syn.: * N. Diet → watery diarrhea * Temporary lactos intolerance. <u>TTT:</u> - IV nutrition - Avoid Cow Milk

بجول هام بدأ بدأ نظري وشفوي وكلينيك

<i>Disease</i>	<i>C/O</i>	<i>Mode</i>	<i>IP</i>	<i>Inf. Period</i>
<i>Measls</i>	Measles V. (RNA)	1- Droplet "Case" 2- contact é contaminated articles From respiratory Secretions of case.	1-2 w	Through out Dse 10d
<i>Rubella</i>	Rubella (RNA)	1- 2- 3- Transplacental	2-3 w	Last w of IP & 2w After Cong. Rubella à 2y
<i>mumps end. parotitis</i>	Mumps v. (RNA)	1- 2-	2-3 w	1d à swelling à 3d after subside
<i>Chicken box "varicella"</i>	varicella zoster v. "DNA"	1- 2-	2-3 w	1d à rash à 7d
<i>Diphtheria</i>	C. Diphtheria	1- 2-	< 1w	
<i>Whooping Caught</i>	Bordettela pertusis	1-case no carrier .	1-2 w	
<i>Scarlet fever</i>	gp A β hemolytic streptococi	1-Case, Contact 2- Mixed born inf	< 1 w	
<i>Poliomyelitis</i>	<u>Polio v. RNA</u> "picorna v. group" I- epidemic II- endemic III- sporadic	1- à iso 2w 2- Feco-oral à iso 8w	1-3 w	

Viral

	Measles	Rubella	Mumps	Chicken Pox
c/p	Prodroma - FAHM - mm. à catarrhal inflammation . - ☺ Koplik spot ☺ pathognomonic - Grayish white mem. On erythematous background - Opposite 2 nd molar tooth	- LN: Cervical , Periauricular Occipital pathognomonic	- Pain behind ear on swallowing & chewing	
Prodroma				
Stage	Rash - in 5 th day à last 5d - MP - behind ear à face, Neck , trunk , - Fever↑	Rash - MP - Face à L.L in 24 hr. - inf à solid immunity - Fever ↓	Salivary gland Enlarge * Parotid : - push ear lobule for upward - tender - cover é oedema - skin over normal * Sub (ling. mand.) : rare	Rash : - vesiclopapular - centripedal esp. back - itching هرش - drops on erythm. Base
Conv.	- Improve symp. - Branny desquamation - Skin Discoloration	No desquamation	- After swelling subside	Crust fall é out scar
D. D	AE of MP rash	STORCH	- Parotid swelling - cervical LN - Dental condition	Papulo-vesicular rash
inv	- V. Isolation : BL , urine - ↑ # meals AB - CBC: leucopenia relative lymphocytosis	- V. Isolation : BL - IgM For rubella - history maternal rubella during pregnancy	- V. Isolation : BL & CSF - leucopenia , relative lymphocytosis.	à Fluid of vesicles
Prophylaxis	A à Measles v. , MMR P à gamma globulin	A à MMR vaccine P à Hyperimmune	A à MMR P à Ig	A à Live attenuated P à zoster IgG

	NB: <i>A = Active</i> <i>P = passive</i>	serum		
TTT	<u>General</u> - isolate 1w` - Bed rest - Antipyretic & analgesic - Good diet.	- But no isolation - Avoid contact é Pregnant female	- iso. Till swelling disappear - Antipyretic ,analgesic - Diet : no sour	- bed rest - light meals - Itching: calamine ; sys. Antipyretic
	<u>Specific</u> - gamma globulin - Vit. A		No specific	Acyclovir # v.
	Comp. 1. AB à 2 ^{ry} inf 2. Anticonvulsant à Epilepsy 3. Correction of dehydration à Diarrhea			
Complications	<u>CNS :</u> - Hemiplegia - Meningitis – Encephalitis - : After 5 d of rash <u>A/E:</u> 1. V. invasion 2. Ag/Abreaction <u>Ear:</u> - O.M - <u>Cervical L.N</u> <u>Eye</u> Keratitis – conjunctivitis <u>CVS :</u> Myocarditis Respiratory : Rhinitis – Sinusitis – Bronchitis – B.pn <u>Skin:</u> Purpura – hemorrhagic rash – bleeding mm. – infection : impetigo , frunculosis	<u>CNS :</u> Encephalitis <u>Bl:</u> Thrombocytopenia <u>Joint:</u> Polyarthritis <u>Congenital Rubella Syn</u> <u>A/E:</u> - Transplacental spread. - Newborn discharge viruse in his secretions 18 m` " infective " <u>C/P :</u> <u>Early:</u> <u>Brain</u> MR,C.P ,Microcephaly. <u>Bl:</u> Thrombocytopenia <u>Bone:</u> ↑ area of translucency . <u>CHD:</u> VSD, ASD, PDA. <u>Lung:</u> pneumonia <u>Ear:</u> Deafness <u>GIT :</u> HSM <u>LATE:</u> Brain : MR Eye: Blindness Ear : Deafness. <u>INV:</u> Hx of rebula during preg., V .isolation. (Bl) <u>DD :</u> TORCH Prevention: MMR , H.I.S	<u>CNS:</u> Meningitis , Encephalitis, facial palsy <u>Pancreatitis</u> C/P: Acute Abd INV: ↑↑ leukocytosis S.amylase <u>Hepatitis</u> <u>Myocarditis</u> <u>Orchitis</u> Painful swollen tender,unilateral <u>Oophoritis</u> Abd. Pain Thin tunica albuginea	<u>CNS:</u> Encephalitis, facial palsy <u>Eye</u> Keratitis – conjunctivitis <u>CVS :</u> Pancarditis Lung: pneumonia Neonatal varicella Mother infected 5 d befor delivery → Baby Death Congenital vaicella mother infected in 1 st gestational 20 w` → Microcehaly LBW

-

	<i>Roseola infantum</i> (<i>Exanthema subitum</i>)	<i>Erythema infectiosum</i> (<i>fifth disease</i>)
<i>Organism</i>	Human Herpes 6	Parvovirus B19
	Infant (6m - 2 y)	School children.
<i>Incidence</i>	Droplet	
<i>Mode Of Trans.:</i>	I - Prodroma	
<i>C/P</i>	- 4 days - High Fever (39 – 41) without any localizing sign (irritability , convulsion , bulging AF , no cough , Conjunctivitis , rhinitis).	No Prodroma
	II – Diagnostic stage	
	- Onset : 4th day of fever - Relation to fever : temp. Drops rapidly as rash appear " Rainbow follow storm " - Type : Maculopapular non pruritic. - Site : trunk - Spread : arm , neck , with minimal face involvement - Final distribution: fade in 24 hrs. - No desquamation or pigmentation.	- Last for 2-40 days - pruritic - Resolve without desquamation - Recure with exercise, warm bath, rubbing skin ,emotional upset . - passes following stages : 1. Erythema facial flushing : (slapped check appearance) with circumoral pallor. 2. Maculopapular rash : of trunk & extremities (extensive surface then flexors). 3. Central clearing of rash : giving a lacy appearance .
<i>IP</i>	1 – 2 weeks	1 – 4 weeks
<i>Investigation</i>	Leucopenia with relative lymphocytosis.	- Normal WBCs - IgM to virus during 1st month after onset of illness.
<i>Complications</i>	د / ماح & د / الجيزي	1. haemodynamic Anemia . 2. Arthropathy 3. Encephalitis . 4. Infection: Foetal: IUFD, hydrops immunocompromized (anemia). 5. Pneumonia.
<i>TTT</i>	No specific ttt	
	- Antipyretic. - Sedative : For Convulsion	- IV Ig – Isolation Of Contact with hemolytic anaemia

Bacterial

	<i>Diphtheria</i>	<i>Whooping cough</i>	<i>Scarlet fever</i>
<i>AE</i>	Cornibacterium Diphtheria	Boardeltela Pertussis	gp A B hemo. streptococi
<i>Mod</i>	- Droplet - Contaminated Articles	- Droplet "Case only"	- Droplet (case – contact) - Milk born infection
<i>IP</i>	< 1 wk	1-2 wk	< 1w
<i>C/P</i>	- <u>According to the site</u> 1 – pharyngeal : - Fever – headache - dysphagia – sore Throat . - CV. LN enlarge Tonsil Congested , enlarged Covered é mem :- White or dirty gray (RBC) adherent IF removed à bleed 2 – laryngeal : AE : $\frac{1}{3} \rightarrow 1ry$ $\frac{2}{3} \rightarrow 2ry$ 2 ph. - fever Hoarseness - Steroidor - suffocation 3 - Nasal : - Serosagenous discharge - Excoriation of upper lip. 4 - Vaginal , cutaneous Ulceration , mem. Formation . 5 - conjunctival : Injection , mem. formation	1 - Catarrhal stage:(1-2 w) Fever- conj. Injection – lacimation – rhinorrhea 2 - Whooping caught : - 2-4 w - Caught : – paroxysmal 5 -10 Caught followed by inspiration against narrow glottis à whooping <u>Ass. e :</u> Facial redness – cyanosis – lacrimation – salivation – protruded tongue – distended N.V , vomiting ☺ Caught & vomiting à pertussis ☺ - triggered by sneezing yawing , eating , drinking 3 -convalescence (1-2 w): - ↓ severity , no whoop. - child exam :- Rhonchi, consonating crepitation	Prodroma: Acute onset : Fever, vomiting, sore throat Throat: - ant. Pillars oedema - tonsillitis é or é out yellow membrane - hard palat à red sports Eruptive Stage : After 12-48 hrs – P. areas - diffuse Erythema P - Fine maculopapulse à goose skin. - fade on p. but transverse Line appear"pastia's line" Tongue: While é red points " white straw berry " After 7 days: - Rash à fade é desquamation - tongue à swollen-red "red strawberry tongue"
<i>DD</i>	<u>AE of mem. on tonsils:</u> امل A Acute tonsillitis M Moniliasis L Ludwig's angina	<u>AE of paroxysmal caught</u> B-pertusis – Adenovirus <u>AE. Of signal wheeze :</u> - Acute bronchitis - FB à A. onset	<u>AE of MP rash.</u>

-

		- Bronch. Pn à chest signs. Laryngismus Stridulus à <i>tetany</i>	
Lab	- +ve mem. swab à diph. - +ve culture on loffler's serum . - ECG : Arrhythmia . T wave changes . - WBCs : ↑ . - Urine , albuminuria . Hyaline casts .	-Leucocytosis : 20000-50000, relative Lymphocytosis . - ESR : ↓ . - +ve N. ph swab . - +ve culture on BordetGengou Media . - Serology	- Leucocytosis - Anemia - ASO > ↑ 250 Todd's Unit . - + ve culture thought Swab . - Dick test. - Schultz Charlton test
C/O	1st wk : - circulatory collapse . - Res. Comp: (L.obstructn, Br. pn) 2nd wk : - Toxic Myo – HF- HB 3rd wk : - nasal (regurgitation – Tone Of voice) - Loss (uvular mov. – gag reflex) 5th wk : paralysis : * Extra ocular à squint * Intraocular à loss acomdation 7th wk: vagal neuritis à -HF à sudden Death - RLN à suffocation - Phrenicn à diaph. 12th wk: - <u>motor</u>: tender calf ms - <u>Sens</u>: glove, stocking hypothia.	Res - Emphysema, empyema , pn.thx, bronchopneum., bronchitis - GIT : - <u>Tongue ulceration :</u> dt billing during attack. - <u>Server vomiting :</u> Dehydration, malnution - Hernia, rectal prolaps: CNS - Cerebral anoxia à convulsion - ↓ ca à teteny - ICH - Encephalopathy CVS - Epistaxis - Hemopsis - H.F	Immediate - OM – pyoderma, - sinusitis, - pertonsilar abscess - Larygitis - Born.pn - Emphema - Septesemia - Mengitis - Arthritis Delayed - AGN - RH fever - Henoch Schonlln purpra "Anaphlactoid purpra"
Pro ph	- DPT - Patient : isolation - Contact : naso ph. Cuture If : -ve : vaccinenation	Active : DPT , P. vaccine toxiod Passive : hyperimmune pertusis gamma globuline	

-

	+ve : isolation , AB till 3 succive –ve cultures.	Chemoproph: Erythromycin, ampicillin 10d	
<i>ttt</i>	General : - Isolation bed rest (1 m) - Good diet Specific : antitoxin 1M-1V 10,000 - 100,000 IV AB : pencillin – erth. ttt Comp. - Myocarditis : diuretics , digoxin - Circulatory .f : BL. Trans. - neuritis: limb splitting – physiotherapy	General : - Rest., isolation - O2 , section Removal, - diet , light IV fluids - sedation, anticonvulsant - Antitussive à caught Specific : Ampicillin- erythromycin eliminate organism in 3d. ttt of Comp.: <u>N.B: MCQ</u> All ages R susibtable No passive imm. No transplacental imm.	General : - Bed rest - Antipyretic - Analgesic - Diet à ↑ fluid , calories Specific : - <i>Procaine pencillin</i> 1m for 10 days 600000 1u 1d à of choice followed by <i>LA pencillin</i> if sens. Use erythromycin . TTT of complications

Polio myelitis

AE : Poliovirus "RNA"

Type : I : epidemic II : endemic II : sporadic

No cross imm. لو الطفل اتعدى بياخد التطعيم بردو

Essential For D :

- LMNL (↓T, ↓R, degeneration, Disuse atrophy)
- Purely motor .
- Asymmetrical Distribution .
- Stationary or regimen coerce .

Mode of trans :

- Direct droplet
- Fecooral

IP : 1-3 w

Pathogenesis :

Pharynx à payer's patches & 1N of intestine à Blood. à viremia if not neutralized é Ab. à CNS paralytic manifestation.

-

C/P **Asymptomatic 95 %**

virus found in throat, faces.

Abortive

Acute Febrile illness é

Fever not exceed 39 .5 C

Headache – nausea – vomiting- abd. Pain , constipation, URT, sore throat & cough

2/3 of clinical cases

Non paralytic

1st phase : As Abortive.

2nd phase :

1- meningeal irritation.

3- ms rigidity.

2- nuchal rigidity.

4- Impending paralysis

1- Meningeal. Irritation

A. Burdzinski sign :

é ptn. Supine , LL. Extended

flexion of 1 LL. à flexion of 1 other.

B. Tripod sign :

Ptn. à Can't sit é out support.

à if sité .. é hand behind

NB. A,B Disappear in prone position.

2- Nuchal rigidity signs:

A. kernig's sign

Inability to extend knee é spasm of hamstring ms. When thigh flexed at hip. At night angel.

B. head drop sign:

hands under ptn. Shoulder ,raise trunk à head drop back.

3- generalized ms weakness , tenderness

Ex. In extension of LL.

-

4 - signs of impending paralysis

- Ms weakness
- Reflex either ↑ or ↓
- Loss of :
 SuP. Reflexes : spinal, aemastic, gluteal
 Deep Reflexes: afterwards

Paralytic

Non paralytic & weakness of one or more of spinal or cardinal MS.

Types :

Spinal à *lumber* : L.L , L. Abd.
 à *Thoracic* : intercost , U.V
 à *cervical*: shoulder, arm,
 diaphragm

Bulbar :

Paralysis of 9,10,11 à pharynx, tongue, larynx .
 involve of vital centers à
 R.C HRC C.C

Bulbospinal : Mixed

Encephalic : Drowsy
 irritable
 tremor

D.D. of polio like Acute Flaccid paralysis**1-Non Paralytic :**

Meningitis "Septic - aseptic"
 Meningism é pneumonia
 Rheumatic Arthritis

2- Paralytic :

1- *gullian barre syndrome*.

Symmetrical – sensory affection .

2- *post-encephalitis*.

3- *peripheral neuritis*.

Symmetrical – sensory affection .

4- *transverse myelitis*

Symmetrical - Sensory loss .

5- *pseudo paralysis* : Painful condition in joints à limited mov. eg. Synovitis .

-

6- tick bite : improve after remove tick .

7- post encephalitis : UMN , spasticity .

8- Post Diphtheric :]symmetrical sensory affection - sore throat

3rd wk à N. regurgitation

5th wk à squint

Complication

1- **Res. Failure** à wet : bulbar type "Central respiratory failure "

à Dry : spinal type " Peripheral respiratory failure "

2- **CVS** : ↑ or ↓ BP - arrhythmia

3- **UTI**: urine- retention – calculi- infection

4- **GIT**: gastric dilation .

5- **SK ms**: - Quadriceps : Absent tendon R.

- Back ms: can't sit

Prevention

- Passive : Human Ig IM 0.2mg / kg

- Active : - sabine oral

- salk parenteral

TTT

Abortive : Bed rest , analgesic

Non Paralytic: bed rest , analgesic , Hot fomentation (for pain, spasm)

Paralytic : prostagmine 1mg/ kg / d

-Hospitalization - Sand bags ,

-Avoid IM injection - Hard bags

ttt of Complications

- Respiratory Ms paralysis à ventillator

- Bulbar polio

- Nutrition à IV route

Active polio	Chronic polio
3w	> 3w
Ms tender	No Ms tender
fever	No fever
No ms wasting	ms wasting

-

Stages "D Of Ms Weakness "

- 0 à No contraction.
- I à Contraction
- II à Mov. Not Anti - gravity
- III à Mov. Anti- gravity
- IV à Mov. Anti- gravity & Anti- mild resistance
- V à Mov. Anti- gravity & Anti-resistance

- D
- Acute or chronic?
 - Paralytic or not ?
 - Lumber, thoracic or L. Th. ?
 - Complicated or not. ?
 - Which limb ?

- PPF
- IM injection
 - Cortisone
 - Tonsillectomy
 - Ms exertion
 - Pregnancy

- INV
- CSF examination
 - Complement fixation
 - V. isolation "Throat - stool"

Pyrexia of unknown origin

Def : * fever > 38.3 c°

* More than 3 wk

* Unknown D after 1 w of intensive study in hospital

Causes:

1) infection :

à Bacterial

* TB à UT

à Extra Pulmonary

* -ve x-ray , sputum exam.

* Need

- LN Biopsy in TB Lymphadenitis

- Fundal examination :

Choroid tubercle

* IE à -ve BL. Culture if patient receive AB

* UTI à need urine analysis culture U/S

Other à pyelonephritis ., typhoid

à viral : CMV – IMN

à Protozoa: amebic abscess.

à Fungal : moniliasis

2) Infiltration :

à Neoplastic

- Bl . -Leukemia - Lymphoma

- Bowel - hepatoma -hypernephroma cancer (stomach ,colon)

3) Immune:

SIE Rh. fever Rh. arthritis

4) intestinal :

ulcerative colitis

crhon's Dise .

sarcoidosis .

5) idiopathic :

unknown causes.

6) à Other :

à fever : FMF

à medicine . liver cirrhosis

. multiple pulemboli

à 2 phycogenic :

Habitual & hyperthermic Female

C/p Headache

Insomnia

Fatigue

Diagnosed by exclusion

Factitious :

Fever : false elevation of temp.

By putting thermometer in Hot liquid

D : no rigor

Diagnosis

à History : contact é TB case

Travel to endemic areas

à C/P :

Symp. : chronological develop of Symp .

Sign : general : skin lesions

Cvs : exam

Chest : exam

GIT : exam

GUT : exam

à Inv

1) lab à for AE

- CBC infection

neutrophilia à B inf.

Lymphocytosis à TB

Esiophilia à parasite

- Culture : B1 – URINE

- CRP –ESR : non specific

Immune à ANA in SLE

à Rh. factor in Rh. arthritis

2) radiological :

- Cervical x-ray , Abd. U/S à

Tr . & Abscess

TTT: Medical : therapeutic trial

INH à TB

Aspirin à Rh.artheritis

Metronidazol à hepatic Amebasis

Chloroquine à malaria

4) surgical: *exploratory laboratory*

after failure of all methods of investigation to see Abd. Tr , Abscess

Haemophilus influenza infection

Causative organism:

Haemophilus influenza (6 types a- f) à most virulent is b sero type .

Mode of transmission: droplet .

IP : variable .

Risk group :-

children with à sickle cell anaemia
à asplenia .
à Immune deficiency. malignancy .

C/P :-

may result in :

- 1- meningitis .
- 2- cellulitis .
- 3- epiglottitis.
- 4- Pneumonia
- 5- Bacteremia .
- 6- Others as : UTI , endophthalmitis , pericarditis , osteomyelitis,
otitis media , conjunctivitis or sinusitis .

Dr.Madeh Dr.AlgizY

D :-

- 1) **CBC** à leucocytosis .
- 2) **bacteriology :** à smear stained with gram stain or methylene blue .
سمير زرع سيرو à culture (chocolate agar)
à serology .

Treatment :

- à 1st line :- augmentin , azithromycin , 3rd generation cephalosporin.
- à less effective à ampicillin , amoxicillin , chloramphenicol , cotrimoxazole .

- *FEVER WITH M.P rash*

<i>Lesion</i>	<i>C/O</i>	<i>Rash</i>	<i>Pathognomonic signs</i>
<i>Measles</i>	Measles virus	See Measls	Kolik's spots
<i>Rubella</i>	Rubella virus	See Rubella	Lymphadenopathy esp .post auricular , Occipital
<i>IMN</i>	EBV	- In 10-20% of cases à () 4 th & 8 th day . - Appear as discrete macular , petechial , morbiliform & Scarletiform.	<u>Triad of</u> - memb . tonsillitis , - L.A - splenomegally .
<i>Roseola Infantum</i>	Human herpes Virus type 6	See Roseola Infantum	Rash appear as tear drops .
<i>Erythema infectiosum</i>	Parvovirus B19	See erythema infectiosum	Slapped cheek appearance “ Butter fly “ rash on nose & cheek
<i>Scarlet Fever</i>	GABHS	See scarlet fever	- Strawberry tongue - pastia line - follicular or membranous tonsillitis desquam. Esp. hands & feet
<i>Meningo-coocemia</i>	Meningococci	Popular ,MP , petechial or hgic - no specific distribution	Evidence of meningeal irritation & ↑ ICT
<i>Kawasaki Disease</i>	Collagen dose	Acute phase ch. By : - fever, conjunctivitis ,stomatitis - rash à M.P or scarletiform - edeme of the hand & feet. - CX L.N - myocarditis & coronary aneurysm	

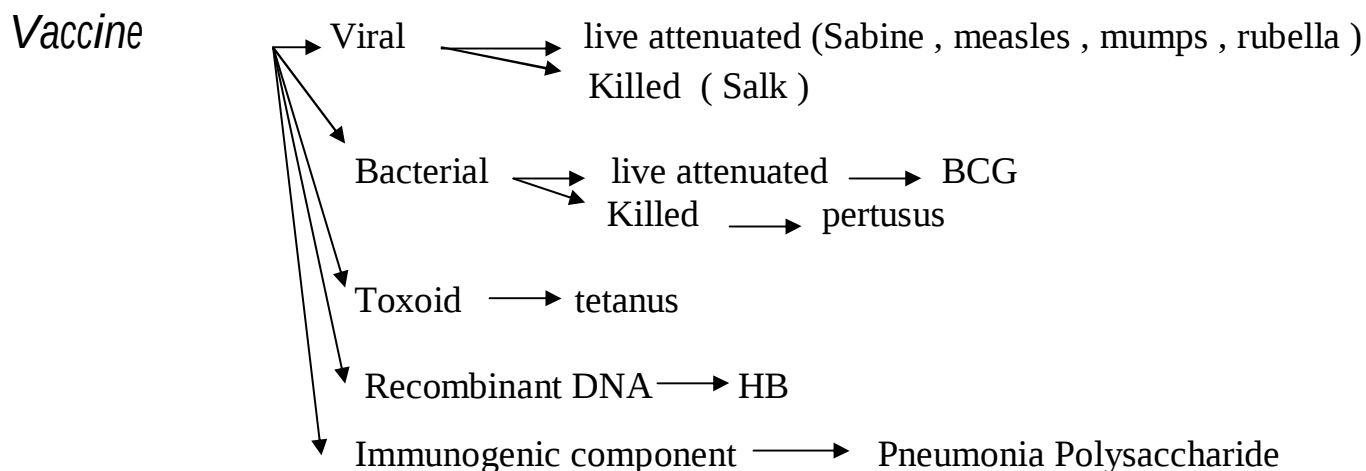
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FEVER WITH P.V RASH

<i>Lesion</i>	<i>C/O</i>	<i>Rash</i>	<i>Pathognomonic signs</i>
<i>Varicella</i> <i>"chicken pox"</i>	V-Z virus	See chicken pox	Pruritus , rash characteristics.
<i>Herpes zoster</i>	V-Z virus	- along dorsal never root course. - with vesicles grouped together. - vesicle → pustules → dry → Scab → clear in 10- 14 day - regional L.N enlargement.	Characteristic distribution & characters. A long dorsal n. Root
<i>Small pox</i>	Small pox virus	- Red macular → vesicular - vesicles → umbilicated , involve all m.m.	Rash Characters
<i>Entero-Viral infections</i>	Enterovirus "echo & coxsackie"	Rebella like, discrete, generalized. Non pruritic no desquamation.	Aseptic meningitis with rash
<i>H, F, & M. disease</i>	Coxsackie –A virus	Vesicles on hand & feet	-Stomatitis with vesicles over buccal mucosa plate, gum, tongue. - Assoc. with G.I. symptoms.
<i>Staphylococcal infection</i>	Staph.	- Generalized erythematous, scarlatiniform. - Sand–paper texture, tender skin - Bullae appear with in 2 days & epidermis separates into large sheets, revealing a moist red surface	Associated staph. Skin infections e.g. impetigo or purulent conjunctivitis .

Immunization

Immunization → active: induce immunity by vaccination : (vaccine , toxoid)
 → Passive : transplacental , Ig administration



Schedule

< 3 M`	BCG		
2,4,6 M`	OPV	HB	DPT
9 M`	OPV	Measles	
18 M`	OPV booster	MMR	DPT booster
Vit A	At 9 m 100,000 IU		
	At 18 m 200,000 IU		

- C.I**
1. Very
 - sever illness
 - High fever
 2. DPT → IF reaction to it → use DT instead
 3. BCG in → HIV , immunosuppressant

S.E → **LOCAL ASPR** (Abscess – Swelling – Pain – Reaction)
 → **SYSTEMIC** (fever – rash – allergic reaction – convulsion)

CASE (1) 24 m` Baby Never Take Vaccine

24 m	BCG	MMR	OPV	DPT	HB
25 m			//	//	
26 m			//	//	//
27 m		Measles	//	//	
29 m					//

Vit A at 24 m , 28 m

CASE (2) : Premature :

As full term but HB vaccine at 1m` old when wait < 2 Kg .

	POLIO		DPT	MEASELS	MMR	BCG
TYPE	SABIN Live attenuated	SALK Killed	Diph. , tetanus, pertusus Toxoid	Live attenuated	Live attenuated	Live attenuated
DOSE	4 drops	1 ml	0.5 ml	0.5 ml	0.5 ml	0.05 ml at 1m 0.1 at 6y 0.1 at 13 y
ROUTE	Oral	S.C IM	Deep IM	IM	18 , 24 m	Strict ID Lt deltoid
Indic.	2,4,6 m 1.5 , 6 y	2 nd m	2 ,4 ,6 m & 1.5 , 6 y	9 m		ABOVE
C.I	Advantages : * Cellular , Hormonal immunity * Cheap , easy * Solid immunity.			Fever Pregnancy ↓ Immunity Cancer		Others : Phenomena after vaccination 3w → 5w → then Papules 8 mm subside
S.E		Disadvantages : * Only sys. imm. * Not use in epidemic * expensive	<u>Mild</u> : F S T Fever - swelling - tenderness <u>Moderate</u> .: Shock like <u>Sever</u> : Convulsion Brain damage Encephalitis	Fever Rash Encephalitis convulsion	Arthritis	

<i>vaccine</i>	<i>Pneumococcal. Polysaccharide</i>	<i>Meningococcal Polysaccharide</i>	<i>HB</i>
DOSE	0.5 ml	0.5 ml	2,4,6 m and 1.5 y
ROUTE	S.C	S.C	
INDICATION	Asplenia ↓ Immunity DM	CLOSED COMMUNITY	

Rheumatic Fever

Def : diffuse inflammatory Dse. of C.T. of Ht , Joint , CNS ,S.C tissue .

AE : delayed sequelae of group pharyngeal infection e' gp A β hemolytic Streptococci

Pathogenesis : theories :

- 1 – Infective
- 2 – Toxic
- 3 – Auto – immune :-

Evidence :

- 5w () infection & physical finding
- Spontaneous improvement .
- Response to Cortico-Steroids.

Theories :

- * Cross reactivity : Ag of strep, Ht, joint are similar Immunological so Ab formed Against strept. react e' Ag of these tissues.
- * Altered Antigenicity: Strept Alter Ag of These tissues

PPF

الشخص

- M = F (chores $\uparrow$ inf)
- 5 – 15y` ( peak at 8 y`)

عائله

- Congenital
- Genetic factors
- Poor housing conditions

مجتمع

- low socioeconomic status

مكان

- Tropical .subtropical

زمان

- cold months

أهم حاجة

- recurrent strept infection :
 - . most imprtant factor .
 - . may be - Tonsillitis
 - Sever throat infection

Pathology

1 – Exudative charges:

Edema ; cellular infiltration
(Lymphocytes – plasma cells)

A – heart : Endo: valve edema $\rightarrow$ stenosis
Myo : damage - dilatation
Peri : coverby fibrous exudates

B – joint : - Swelling - synovitis
- Serous - effusion

2 - proliferative changes:

- . pathogonomic " aschoff`s body "
- . perivascular according to multi – nucleated giant cells.

Forms : . Vegetation on valve leaflet.
. S.C nodules.
. CNS. In chorea.

Diagnosis

History :

- + ve family history - Easy fatigability
- Epistaxis - Previous strept
- Pallor - Abd .pain
- Pharyngitis (2 – 50) age.

Ⓣ of acute RH. Fever :

<u>A. major</u>	<u>B . minor</u>
• Polyarthrits • Rh . carditis • Rh .chorea • S.c nodules • Erythema Margenatum	• Clinical: Fever ; arthralgia • Lab : $\uparrow$ ESR, CRP, $\uparrow$PR
<u>Plus :</u> C. <u>Evidense of strept infection :</u> - $\uparrow$ ASO titre - Rapid Ag elevation - Culture	

1. Jones criteria

2 major or (1 major + 2 minor)

2. Revised Jones

- As Jones
- Evidence of strept .infection ... *تذكر*
- + precautions :
 - if arthritis is Major → don't take arthralgia or Fever minor
 - if carditis is Major → don't take ↑ PR as minor.
 - S.C nodule Erth. marginatum : not specihc

3 – WHO recomondation

- we can Ð rh .f e'out 2 major or 1 Major + 2 minor in :
 - chorea e' no other explanation .
 - carditis
- Recurrent Rheumatic Fever .

Major Criteria**1 – polyarthritis 75%**Jonits : large: " knee , ankle , wrist "Character :

- joint is hot , red , swollen , tender , painful .
- Limited movement.
- migratory.
- spontaneous improvement in 10 d .
- dramatic response to salicylate

N.B : Arthritis means :

- Swelling only Or..
- 2 of : 1. pain . *بطل*
- 2.Tenderness .
- 3. Limit. range of passive movement.

2 – Rh carditis : 40 % - Pancarditis ➡**A _ pericarditis***** Dry pericarditis**

- Symp : chest stitching pain
- Signs : - Pericardial rub
 - To and fru _ sys. Dias
 - ↓ If effusion occune

*** Pericardial effusion**

- Auscultation : weak H.S
- X-ray : ↑ Cardiac shadow.
- percussion : ↑ Cardiac dullness .

B – myocarditis

- Gallop rhythm.
- Tic tack // " no ms component of S1
- Cardiomegally .
- Tachycardia: out of proportion e' fever
- HF .
- ↑ PR
- ↑ CRP , ↑ ESR , ↑ MB

C – Endocarditis

- * Affect :
 - Mitral > Aortic > Tricuspid never pul.

Mitral Valvulitis	
Edema → MS	Destruction → MR
* Mid -diastolic e' presystolic Accentuation	* Pan. Systolic
* Rumbling	* Blowing
* Apex , not prop.	* Apex → Axilla
* Low pitched	* High pitch
* ↑ e' Expiration	* ↑ Exercise, on lt side

N.B: Carry Comb`s murmur : MS murmur in Rh. Fever .

Aortic valvulitis

Edema → AS	Destruction → AR
* Ejection sys	* Early diastolic
* A1 → Apex	* A2 → Ap
* Harsh	* Blowing
* Crescendo	* Decrecho
* Decrescendo	* ↑ e leaning forward

N.B :

Austin flint murmur :Functional MS during AR.

3 – RH .Chorea 10 %

- F > M
- 6 m interval () infection onset Of It.
- Spontaneous improvement e` in 6 m`
- No acute phase reactants.
- Never e' Arthritis.
- **C/P triad**
 1. **emotional liability :**
 - Alternate laughing , crying
 2. **Hyporonia :**
 - Extensors > Flexors .
 - ↓Tonia → paralytic chorea
 - ↓ Reflexia→ pendular tendon jerk
 - Milker maid sign
 - Boat shaped hand → Dinner fork deformity
 3. **involuntary movements :**
 - . Sudden, involuntary, dysrrhythmic
 - . Jerky – semi-purposeful.
 - . proximal > distal .
 - . ↑ e' emotion .
 - . ↓ e' sleep .
 - . **forms**
 - . Grimacing
 - . Stumping
 - . Shacking shoulders, hands.
 - . Indistinct speech

- Tests :

- . **Arm elevation:** failed.
- . **Tongue protrusion :**
 - Maintain only on holding by teeth.
- . **pronator sign :**
 - Unable to maintain supination, Pronatic
- . **piano player sign :**
 - Extended finger wavy movement.
- . **buttoning & un boun ttoning**

- Types :

- . Usual type .
- . Hemi chorea
- . Chorea mollis: paralytic hypotonia, sever hypotonic
- . Chorea gravis " Choric movement " .
- . Maniacal Chorea : " emotion. liabilily " .
- . Gravidorum
- . Serile

4 . subcutaneous nodules

- . **Indicate** : Server attack , bad prog
- . **Nodules** : Small., Firm , rounded , non tender , mobile
- . **Site** : - bony presence
 - Extensor surface of limbs

5 – Erythema marginatum

- . **Site** :
 - Trunk, proximal of limbs, never face
- . **Ch** :
 - Circular, Erythematous, non itchy

Minor Criteria

1 – clinical

- . **Arthalgia** : migratory .
- . **Fever** : low grad – not e' Chorea.

2 – lab

- a. ↑ **ESR**: non specific, not D for activity
- b. ↑ **CRP**
- c. ↑ **Acute phase reactance** :
 - γ globulin, ↑PR interval, leucocytosis

Rheumatic Fever With

↓ ESR	CRP : -Ve	ASO : -Ve
Iso.chorea	Iso.chorea	Iso .chorea
Eryt . marg.	Eryth. marg	
Late onset carditis		Late onset carditis

Evidense of strept infection

- 1- Culture → causative organism
- 2- Ab : - ASO titter > 250 toad unit → D
 - Anti DNA ase
 - Anti NAD ase
 - Anti Streptokinase
- 3- Rapid Ag detection test

Investigation

For strept.	- Culture - Anti strept Ab
For Rh. Activity	↑ ESR CRP + ve ↑ PR
Other	Bl : leucocytosis
	X- ray Cardiomegaly
	ECG - ↑ PR , myocarditis - Elevated ST → Pericarditis
	Echo - sub-clinical

D of Rh . Activity

- Sleep pulse > 100
- Cardiomegally
- H.F.
- New murmur
- Pain
- ↑ ESR
- CRP + Ve .
- S.C nodules
- Fever $\geq 38\text{ C} \geq 3$ days

TTT**1- Bed rest**

	Bed rest	Gradual ambulation
Arthritis	2 w	2 w
Carditis	4 w	4 w
Carditis , Cardiomegally	6 w	6w
Carditis , HF	12 w	12w

2 – Diet : - ↑ Vit C

- ↑ Salt restriction

3 – AB

Eradication	Recurence
Procain p. 600.000 IU 10/ d	phenoxy methyl blue 250 mg / 2 / d
bezathin p. .Single IM . <lt 27y'="" 600.000="" iu<br=""></lt> > 27y` 1.200.000IU	bezathin p . .Single IM . For 3w شفوى
Erythromycine 40 mg / kg /d	Erythromycine 250 mg / 2 / d

*** Never use Tetracycline – sulfonamid**

Bacteriostatic → resistance

4 .anti – inflammatory1 – **Arthalgia** → salicylate (analgesie)

2- **Artheritis , carditis** → salicylate
for 2w 100mg / kg /d
for 2 w 75 mg / kg /d

3- **carditis e` HF , cardiomegaly** → **Predinsone**

- 2mg l kg/d ÷ 2 doses FOR 2 w`
- Withdrawal in 2 w.
- Under cover of salicylate
75 mg /kg/d for 4w to Prevent rebound phenomena

S.E of satycilate

- Tinnitus
- ↑ hyperventilation
- ↓ hypoglycemia
- Metabolic acidosis
- Bleeding tendency ↓ prothrombinemia
- Gastric irritation

TTT of chorea

- A . ↓ stress
 B – plasma exchange & IV Ag therapy
 C – as before penciline & chemoprophylaxis
 + هدفك ..
Halopridol 0.05 – 0.5 mg / kg / d
Diazepam 0.3.....
Phenobarbiton 5
Chlorpromazine .

TTT of HF

- A – gerenal lines
 Bad rest semisetting position .
 Diet : - small frquent
 - salt restriction
 Sedative : diazepam
 Morphine sulphate: 0.5 mg / kg
 B – C-S :
 + Ve intoropic : digitalis
 C – Diuretics : frusmide – spironolactone
 D – V.D : Na Nitroprosside

Prognosis

- permanent cardiac damage depend on :.
 - HT : 1. Stat at the beginning of Dse.
 2. Regression of Dse
 - Recurrence of RH.F.

D.D of chorea

كف طب

C/p

- **Congenital** C F T P
- **Familial**
- **Toxic**
- **Post " Encephalitis , Kernictrus "**

D.D of arthritis

كام ای بی CAM IB

- **Collagen Dse** → SLE
- **Allergic Arthritis**
- **Malignancy** : leukemia
- **Infection** : Salmonella .Shegella
- **BL** . : hemophilia

D.D of carditis

- Common in children
- Pul. → systolic
- parasystolic → musical
- Not propagate
- Change by change i position
- Soft grad I or II
- Short duration
- No charge in H.S

نهاية الدراسة...



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Congenital Heart Disease

	CYANOTIC	ACYANOTIC
<i>RVE</i>	T4 T3 ; Eisemenger Syn.	PS ASD P.HTN
<i>LYE</i>	Tricuslid atresia	AS PDA Coartctation of Aorta
<i>R; LYE</i>	(TA) Truncus Artetiosus (TGA) Trans position of G.V	(BIG VSD) P. HTN

V.S.D

Types :

1 – Acc.to site:

- * Membranous (i commonest) .
- * Muscular .
- * Endocardial cushion defect .
- * Sub – pulmonary defect.

2 – Acc to size :

- * Small (< Aortic Annulus) الدكتور هاله المرفاوى
- * Moderate
- * Large (> Aortic Annulus)

3 – Acc.to significance :

- * Singnificant shunt : Pul : Sys. flow → > 2 : 1
- * Non – signif shunt : Pul : Sys. flow → < 2 : 1

4 – Acc.to.number :

- * Single
- * Multiple (Swiss cheese)

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Hemodynamic

1 – **If Small:** large resistance to Lt to Rt shunt → normal cardiac chambers & pulm v. b

2 – **If Large :** minimal resistance → 1. ↑ pulm . artery blood & pressure

2. lung plethora

3. biventricular enlargement.

- RV (Press. Overload)

- LV (Volume Overload)

C/p

1 - if Small : (Roger`s disease):

Symptoms : no

Signs : * Normal S1 , S2

- * Murmur : pansystolic murmur(Loud harsh or blowing heard over Lt 3rd, 4th parasternal area)
- * Thrill : Propagated all over pericardium .

2 – if moderate : as small +

- Slightly accentuated (S2 → P2)
- Apex : functional MS (mid-diastolic tumbling murmur)

3 - **If large :**

N.B : No low COP in VSD شفوی د هاله المرصفاوی

Symptoms * dyspnea .

- Recurrent chest infection .
- Feeding difficulties & low B.wt .
- Growth retardation .
- HF in early infancy .
- Long standing P.HTN → ↓ activity .

Sings : 1 – insprpalp & percussion:

- Pericardial Bulge (biventricular enlargement) .
- Apex : shifted outward .& downward & Hyperdynamic
- Pulm .aretery : palpable S2 & dullness
- Epigastric area : pulsations (RVE)
- Lt. parasternal : pulsation + thrill

2 – Auscultation:

- Lt. parasternal area : pansyst . Murmur (less harsh)
- Apex : short mid – diastolic rumbling
- Pulm. area : Accentuated P2 (P.HTN) e' narrow spitting .

Fate & C/O

1 - Spontaneous closure : in I first year of life & muscular > mem.

- 2 - Complications :
- | |
|--|
| 1 - Eisenmenger syndrome : reverse of i shunt → cyanosis & clubbing |
| 2 - Biventricular failure 3 – inf . endocarditis. 4 – A. regurge |
| 5 – Arrhythmia 6 – haemoptysis 7 – recurr .chest inf |

Investigations

1 – Chest X.ray : - Small : Normal

- Moder : LVE , LAE , P. HTN .

- Large : LVE , LAE , PVM , RVE , P. HTN

2 – ECG : as x-ray

3 - ECHO : Diagnostic

4- Catheterization: D & indicated in : Atypical Finding & ↑ PVR

Managemen

- 1 – small & moderate :
- * Reassurance .
 - * Follow up for spontaneous closure.
 - * Prophylaxis against IE.

Cardio 7 / 21

2 – large : 1 – medical :

- * ttt of chest infection & H.F .
- * Prophylaxis Against IE .
 - All patients : Amoxicillin oral .
 - Allergy : Erythromycine oral .
 - High risk group : Ampicillin IV + Gentamycin IV .

2 – surgical

- * Indication :
 - No spontanous Closure .
 - Significant defect .
 - CHF in early infancy .
 - PA pressure > 50 % of sys . blood P.
- * Procedures:
 - Immediate surgical closure .
 - Pulm . art . banding (multiple & apical defects) .

Fallot's tetralogy (f4)

- Most common Cyanotic Ht Dse (10 % of all CHD) .
- Cyanotic Ht. dises :

- | | | |
|--|-------------------|------------------------|
| * <u>e' ↓ pulm blood flow :</u> | * TOF | * PA + VSD |
| | * Ebstein anomaly | * PA + intact septum . |
| * <u>e' ↑ pulm blood flow :</u> | * TGA | * Common atrium |
| | * Trunk . A . | * Single ventr + TGA |

dt displacement of infundibular septum .

- 1- puimonaty stenosis : * valvular (10 %) * subvalvular (45 %)
- 2- VSD : * large * membranous * non- restrictire & silent dt equal press usually.
- 3 –RVH : Mild
- 4- Over- Riding Aorta : receive blood from both ventricles

Pathology → As above + Aortic arch : Rt . sided in 20 % of cases .
 → Coronary artery abnotmalities.
 → Venous abnormalities

C/P

Symptoms :**1 – cyanosis :**

- At brith : most patients are symptomatic
- Site : tongue . lips . fingers. Toes
- Infant e' mild PS : (Acyanotic Fallot)
 - * initially → HF . e'out cyanosis .
 - * later → Cyanosis late in 1st year .
- Infant e' server PS:
 - * Neonatal period → Cyanosis

* Closure of PDA → sever cyanosis + circulatory collapse .

2 . squatting position : prefer it because :-

- Kinking of popliteal & inguinal arteries → bl .press in aorta → shunt of blood. from R.V. to aorta & force it to pass through stenosed PA → lung oxygenation → hypoxia ↑ LL. pteasure : ↑ cerebral blood flow .

3. symptoms dt hypoxia :

- Dyspnea : on exertion .
- Clubbing : of fingers by age of 1 – 2 yrs .
- Fatigability : easy
- Feeding difficulty → poor growth .

4 – cyanotic spells :

Def : sudden attacks of ↑ cyanosis , dyspnea , irritability or syncope due to more ↓ of already ↓ pulm. bl. flow

AE : 1. spasm of Infandibulum : dt cyanosis & hypoxia

2. systemic resistance : excecise , stress .. fever , infection , ↓ Fe , ↓ Fluid

C/P :

- 1 – Onset : spontaneous unpredictable in the morning
- 2 – Duration : few mins. → hrs
- 3 – ↑ cyanosis & dyspnea & irritability
- 4 - Level of consciousness .
- 5 – Acidotic respiration (rapid deep)
- 6 – Murmur : disappear or ↓ intensity

Invest : ABG : - Hypoxia

- Hypercapnia
- Metabolic acidosis

Signs :

1 – General :

- * pulse & BI . P → Normal
- * R.R → ↑↑ may be present
- * Clubbing → Earliest sign
- * Cyanosis → Central
- * body wt . → ↓ normal

2 – Local cardiac exam :

- * Insp . & pall & percussion :
 - * Pericardial bulge
 - * Quite pericardium
 - * Apex : shifted outward
 - * Lt . parasternal : systolic tutill
- * Auscultation :
 - * Pulm . aera : accentuated single S2 (loss of P2) .
 - * Murmurs:
 - No VSD murmur . هام
 - No murmur during cyanotic spells.
 - Ejec. Sys. murmur dt PS: loud ,harsh ass. e' thrill .

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من أذكار الصباح والمساء :

عن ابي هريره مرفوعاً (من قال حين يصبح
وحين يمسي سبحان الله وبحمده مائة مرة
لم يأت أحد بأفضل مما جاء به إلا أحد
قال مثل ما قال أو زاد عليه) .
صحيح مسلم

- Continuous murmur of PDA & collateral (intense & PS) .

Complications

1- Cyanotic spells اشرح

2- Squatting position اشرح

3- Hypoxia manif. اشرح

4- CNS : brain abscess :

C/P : fever , neurological sign .

INV : CT brin .leucocytosis

TTT : Antibiotics & surgical Drainage .

5- CVS : - HF : rare & never in pure TOF

- CHF may occure : * anemic hge * mild PS & VSD

- Inf.endocarditis :

Site : on Infundibulum of pulmonary valve .

C/P : اشرح

Inv : اشرح

ttt : اشرح

6- Chest : pulm.TB: Cause : lung oligemia

C/p , Inv , ttt

7- Blood : Iron deticiency anaemia : C/P , INV, TTT تذكرها

Bleeding tendency : due to : ↓ platelet number , ↓ Pt , Aptt & ↓ factors 5 & 8 .

Polycyathemia : due to : hypoxia → ++ B.M → RBC's

TTT : partial exchange teanst & can be prevented by good hydration .

Thrombosis : due to : polycythemia & dehydration

Site : cerebral veins& dural sinus .

TTT: adequate hydration & exch .transfusion & Heparin

8 – joints : hyperurecaemia .

9 – skin : scoliosis .

Investigations:

1- Chest x- ray : * boot shaped heart

* R.V. apex

* Lung oligemia

2- ECG :

* RVE

* Right axis deviation

3- ECHO :

* RVE

* VSD

4- Catheterization : confirm diagnosis

5- Blood : - CBC (polycythemia & Iron ↓ anemia)

- ABG

من اذكار الصباح والمساء :

عن أبي هريره قال : جاء رجل الى النبي صلى الله عليه وسلم قال يا رسول الله ما لقيت من عقرب لدغتنى البارحة !

قال " أما لو قلت حين أمسيت " أعوذ بكلمات

الله التامات من هر ما خلق لو تضرعت " !!!!
أخرجه مسلم

*Treatment:***1 – Medical TTT :**

* Prophylactic :- Against Infective Endocarditis.

- Against cyanotic spells * propranolol * Fe & Folic acid

* Curative : 1- of polycythemia:

2- of cyanotic spells :

- Position : knee – chest position .
- Propranolol : relax infundibular spasm .
- Phenylephrine : ↑ systemic resistance .
- NaHCO₃ : to correct acidosis .
- O₂ : to correct anoxia .
- Morphine : sedation & irritability .
- Emergency shunt operation : if failed above means .

2 – Surgical TTT : The definitive TTT

- Palliative : Shunt () syst .& pulm. circulation → hypoxia → blalock tussing operation .
- Definitive : - Total surgical correction (in 1st 2 years) i best operation
 - Removal of obstruction to RV outflow .
 - Closure of VSD .
 - Valvotomy of stenotic Pul. Valve.

Infective Endocarditis

Def : infection of pericardium characterized by :

1. Systemic infection 2. Embolic phenomena 3. Cardiac vegetation.

A/E : - SBE : infection & Cardiac Dse .

- ABE : infection & Normal Heart.

Infection : - Common after minor surgical procedures

e.g : Tooth extraction , Tonsillectomy .

- Commonest Organism : streptococcus & staphylococci.

Pathology :

- A. Large vegetations : Organism surrounded by fibrin , platelet & RBCs.
- B. Myocardium: Contain foci of necrosis, surrounded by cellular infiltration.
- C. C/P produced by : - Toxemia
 - Embolization
 - Cardiac damage.

C/P :

- A. General :** " Toxemic " :
- Fever , Marked pallor , Malaise.
- B. Cardiac :**
- c/p of underlying Dse.
 - Toxic Myocarditis .
- C. Nervous :**
- Cerebral embolism.
 - Subarachnoid Hge.
 - Meningitis
 - Encephalitis.
- D. Ocular :**
- Conjunctiva petechae
 - Sudden blindness CRAO
- E. Spleen :**
- infarction , → pain & rub
- F. kidney :**
- Acute Diffuse GN (ADGN)
 - FGN : Hematuria
- G. Extremities :**
- Pale clubbing.
 - Splinter he under nail .
 - NODULES : - *Jenway spot* : painless – hemorrhagic – in palm , soles.
 - *Ossleir nodules* : Capillary endothelial hyperplasia dt Toxemia (tender – intracutenous – cut palm of hinge)

INV:

- BL : * CBC : Anemia , Leucocytosis ,
- * Culture : on 3 successive days - during febrile phase – under Aerobic, Anaerobic Conditions.
- * ESR ↑
- Urine : * Hematorrhea
- ECHO: * Vegetation > 2mm

⊕ : IE suspected if cardiac get fever without Apparrant cause for 1w

TTT

Prophylactic	Surgical
• AB to : - Dental procedures - GIT surgery. - Urine catheterization • correct predisposing cardiac lesion	• indication : - valve replacement - prosthetic valve Endarditis - Resistant strains - After 6 wk of adequate therapy - Repeated Emboliztion
Curative : Bed rest - Nutritive diet - AB	

Innocent murmur

Def : murmur without anatomical or physiological Abnormality in Heart or circulation .

Types	A/E	Site	Ch`' By
Still`s		() Lt parasternal & Apex	Ejection systolic Low grad I , II , III
Pul . ejec . sys	BL Turbulence In pulmonary artery	2 nd Lt I.C.S	Ejection systolic Soft blowing
Neonatal prephral PS	Hypoplasia of pul . a. branches at birth	//	//
Supraclavicular Arterial bruit	BL . turbulence in carotid	//	//
Cerical venous hum	BL . turbulence in Jugular v .	Neck root	Continuous Soft Change e' change position Res . to postural maneuvers
	Innocent murmur	Pathological murmur	
H.S	No	Yes	
	No	Yes	
ECG	Normal	Affected	
Site	Normal	Normal	
Propagation	Pul , apex , lt paradternal	According lesion	
Grades	I , II , III	Any	
Characters	Soft	Harsh	
Effect Of :			
Respiration	↑ e inspiration & ↓ e expiration	Tissue lesion ↑e inspiration	
Exercise & Fever	↑↑	↑↑	
posture	↑ e standing	No MCQ	
	↓ e sitting	No MCQ	
	+ Ve posture maneuvers		

TGA

Incidence : 5% & M : F → 3 : 1

Path.

- Aorta Arise from RV
- Pul .a // // LV
- 1/2 of these infant have PFO or PDA

C/P

History:

ابنى ازرق وبينهج يا دكتور

- cyanosis since Birth
- dyspnea , feeding diff

Exam :

اه فعلا دا ازرق وقلبه سريع وصوته على

- Cyanosis
- Tachypnea
- S2 : single and load
- No mumur → in intact ventricular septum
or murmur of VSD PS ejection systolic .

D : HF زى ال

- **Lab** : Hypo (xemia, glycemia, calemia)
- **ECG** : - RT QRS Axis
- RVH – RAH
- CVH in sever VSD
- **X – ray** : - Cardiomegaly
- ↑ Pulmonary vasculature.
- **ECHO** : - D

Natural History

Infant e` - intact VS → Sickest gp
- VSD → Least

Cyanonic

- PDA → CHD
- Cerebrovascular accident : rare

Management

1 . Medical

- Correct met . acidosis , ↓ glycemia , ↓ ca
- PG E 1 → reopen Ductus Arteriosus
- O2 → for hypoxia

2 . surgical

- Procedures , switch RT , IT sided Blood at 3 levels :
 1. Aerial level (mustard operation)
 2. Ventricular level (Rastelli operation)
 3. Great arteries (Arterial switch //)

COA

- Incidence : 80 % M : F → 2 : 1
(turner 30 %)

- Pathology :

- position : Juxtadudenal

C/P

Symp . infant

- ↓ Aorta sup . by RT side blood in
- PDA , VSD

Asymp infant

- ↓ Aorta supply by LV. Blood .
- Good collatenale () proxymal , distal
- Past of aorta
- 85 % cases e' COA → bicuspid Aortic Valve

Symptomatic**History*** In 1st 6w:

- Dyspnea
- ↓ wt
- Poor feed
- Sign of Acute circulatory shock

Exam :

- pallor
- respiratory distress
- oliguria
- Acidemia
- Differential cyanosis
(only LL dt Rt → Lt shunt)
- weak peripheral puls .
- S2 single – loud
- S3 gallop
- Murmur
 - . 50 % no mur
 - . 50 % ejection systolic over peri.

INV

- ECG : normal or
 - RVH
 - LVH
 - Rt axial deviation
- ECHO : show site , extend of COA .

Management**Medical**

- * prostaglandin E1 : reopen D.A
- * Anticongestion measures : - Inotropics
 - Diuretics
 - O₂

* Ballon Angioplasty

Surgical

1. Resection & end – to – end anastomosis
2. Subclavian flap aortoplasty
3. patch . aortoplasty
4. Aconduit insertion

Indication : . CHF
. Circulatory shock.

Mortality < 5 %

Asymptomatic

Occasionally – pain in legs After walk .

وجع في الرجلين بعد المشي

EXAM :-

- normal growth , development
- Arterial puls in leg Absent or weak
- BP in arm > leg
- S2 Accentuated
- Sys. Thrill → suprasternal
- Ejection click → Apex & Base
- Ejection sys. mur → upper RT sternal border

INV

ECG : Lt Axis deviation (Lt ward QRS Axis)
LVH

X-RAY :

- Ribnotch () 4 – 8 rib .
- Aortic Arch Diltation
- E . shap indentation by barium oesoph.

ECHO : 1- mem. in i P.L Aspect of ↓ Aorta .
2- Bicuspid Aortic valve.
3- Estimate severity of i obstruction.

Management**Medical:**

- dental hygin , proph Anti IE
- watch closely hypertension .
- Ballon Angioplasty

Surgical : **indication :**

1. Hypertension e' large sys . P. gradient = 20 mm hg () arm & leg .
2. Sever ↑ BP e' CHF , cardiomegally
3. prominent gradient develop e' Excise

Procedure :

- 1 - Resection & end – to – end anastomosis
- 2 - Subclavian flap aortoplasty

Mortality :- < 1 %

<u>Complications</u> 1. Renal failure (commonest) 2. Recoarctation <i>Past operative Follow up :</i> 1 – <u>Annual Examination for</u> : -COA Recurrence 2 – <u>Prophylactic Anti IE</u> . 3 – <u>Ballon Angioplasty</u> :if recoarctation. 4 – <u>Systolic hypertension</u> → ttt	<u>Complications</u> 1- paraplegia " S.cord ischemia" 2- Rebound hypertension <i>Post operative follow up:</i> 1 – <u>Annual Exomination for</u> - Hypertetion - BP . diff () arms – legs. - subaortic AS - Recurrence COA 2 – <u>Continue Anti IE</u>
---	--

Systemic Hypertension

Def: - Systolic or Diastolic BP > 95 % for Age .

- High normal : 90 – 95 % .
- Sever : > 95 % by 10 mm hg .

Classification:

	<i>Systolic</i>	<i>Diastolic</i>
<i>Normal</i>	< 130	< 85
<i>High N.</i>	130 – 139	< 85 – 90
<i>HTN</i>		
<i>Stage 1</i>	140 – 149	90 - 99
<i>Stage 2</i>	150 – 179	100 – 110
<i>Stage 3</i>	180 – 209	111 – 119
<i>Stage 4</i>	≥ 210	≥ 120

A/E:

* lry or Essential :

* 2ry to Dse : 90 % dt : 1- Renal parynchomal Dse 2- Renal artery stenosis 3- COA

<i>Renal</i>	<i>CVS</i>	<i>Drugs</i>	<i>Endocrinal</i>	<i>Neurogenic</i>
- GN & PN - Renal vascular Dse. - Congenital anomalies - Renal damage	- COA - ↑ Stroke vol: PDA - AR - A-V fistul	- Stenoids - Cyclosporins - Amphetamine - Sympathomimetics.	- neuroblastoma - pheochromoadoma - hyperthyroidism - hyperparathyrdism - cushing's DSe	- Brain Trauma - Gulian Bar Syn - Tumor

Dr. Madeh Dr. Algizy

More Common causes of chronic

Newborn	RAS	COA	
< 6 y'	//	//	R. parenchyma Dse
6-10 y'	//		//
> 10 y'	1ry		//

DHistory FH , AE* **CVS** : COA , Headache , Palpitation* **Drugs** : Steroid – Amphetamine* **Renal** : UTI , Radiation , Trama* **F.H** : Essential ↑ BP, Atherosclerosis* **Endo.** : MS weakness
, Hereditary Rend DsePhysical Examination :Of ↑ BP :

- Infant → Crying – Irritability
- Child → Headache – Dizziness

Of comp :

- Hypertensive encephalitis
- Retinal Hge
- Hypertensive HF
- C/P of underling Dse (2 ry)

Inv :* For renal disocders :

- RFT urine analysis U/S
- Renal angiogram

* For endocrine case :

- Thyroid, parathyroid Hormone assay
- Ca assay
- Mineralocorticoid assay .
- CT - MRI - FOR TH.

* For cardiac cases :

- ECG & ECHO

TTT1 . Essential

- nonpharmacological :- ↓ wt , salt ↓ avoid smoking .
- Pharmacological :-

Diuritics	BB	V .D
Furosmide	Atenolol	Hydralazine
2	2	2
Mg/k	//	//
2/d	2/d	1/d

2 . 2ry

- ttt of underling cause .
- ttt Hypertensive crisis .
- Hypertensive Encephalopathy .

Diazoxide	4 Mg / kg	IV
Hydralazine	0.4 Mg / kg	IV
Frusmide	1 Mg / kg	IV

- Fluid balance :
- Seizure 02 diazepam IV . , When controlled shift oral .

H.F.

Def : Inability of myocardium to deliver adequate COP .

A/E : * Diastolic over load:

- VSD PDA ASD
- Valve regurge
- Single (A – V)
- Hyper (Met – ventilation)

* Systolic over load:

- Sys . HTN
- Pul HTN
- Sever COA
- Sever AS
- Sever PS

* Myocardial Dysfunction :

* Myocarditis (Toxic , Bacteria, Viral)

* CMP (idiopathic – Met)

* RH . F

* Malignancy – hemosiderosis Durgs

* Rhythm dysfunction:-

Sever tachycardia – bradycardia

Causes according age (MCQ)

Fetal :

- CHD
- HB
- Sever anemia

Neonome:

- CHD
- Asphyxia
- Fluid over load

Infant:

- CHD
- Viral myocarditis
- Met (CMP) cardiomyopathy
- Acute HTN

Child:

- Acute HTN
- RH . F
- Viral myocarditis
- Cancer ttt

C/P :-

A . In child

↓ COP

SYMP	SIGNS
- Easy fatigability	- Pale cold skin
- Dizziness, giddiness	- Preph . cyanosis
- Blurring vision	- Pulse , rapid , weak
- Syncope, oliguria ,	- ↓ Bp
- Angina	

Pul.congestion :

SYMP	SIGNS
Dyspnea in exertion	- Dyspnea
Cough → Hemoptysis	- Tachypnea
Roc . Chest Inf	- Bi. fine crepitation
Weezy chest	- Wheeze

Sys . Congestion

SYMP	SIGNS
- Insomnia	- Puffy eye lid
- Headache	- Congested NV
- Oliguria	- Hepatomegally
- Rt. hypoch.pain	- Splenomegally
- Dyspepsia	- Oedema L.L
- EPI pain	
- Ascitis, Oedema LL	

B . In infant

SYMP	SIGNS
- Feeding Difficulties	<u>Traid :</u>
→ ↓ wt gain	- Tachycardia Gallop
- Cough – dyspnea	- Tachypnea
- Irritability	- Retraction
- Restlessness	- Hepatomegally
- Noisy breathing	- Cardiomegally

D

History:

<u>in infants</u>	<u>children</u>
- Poor feeding	- Easy fatigability
- Tachypnea	- Shortness of breath
- Poor wt gain	- Puffy eyelid
- Forehead : cold sweat	- Swollen feet

INV:-

- **X – ray :**
 - Lung congition - Cardiomegally.
 - Pul . oedoma
 - ECG :- Det. Defect type
 - ECHO :- // cause of CHD
 - Asses LV Fuetion .
- **ABG , Electrolytes :-**
 - Hypo - Glycemia
 - Hypo - Calcemia
 - Hypo - oxia

TTT :-

- **Elemination of causes** e.g ↑ BP .
- **Elemination of PPF** e.g fever , infection .
- **Control of HF :-**

A. General measures :

- Cardiac chair or infant seat : relife Res. Distress
- O₂ (Humified 40 – 50 %) → relief Dyspea ,

Cyanosis

- Sedation for 2 days :
Morphine sulfate : 0.2 mg / kg every 4 hrs
Phenobarbital : 2 mg / kg every 8 hrs

B. Drug therapy :**(1) Diuretics****1 – thiazide**

* Chlorothiazide: Oral 30 mg / kg Divided on 3 doses .

* hydro Chlorothiazide: Oral 3

- Act on loop of henle .
- Of choice

2 – loob diuretics

* Frusmide :IV 1 mg/kg/d in 3devided Doses .

* Ethacrynic a . IV

3 – aldosteron antagonist

* Spironolacton Oral 3

SE of diuretics:

1. ↓ K → ↑ dig . tox
2. ↓ Chloremic alkalosis → ↑ digitalis Toxicity

(2) Inotropic Drugs :**A – Digitalis :- Oral dose for CHF**

Age	TDD	Maintainance ttt
Premature	20 mg/kg/d	5 mg/kg/d
Neonate	30 mg/kg/d	8 mg/kg/d
< 2 y'	40 mg/kg/d	10 mg/kg/d
> 2y'	40 mg/kg/d	10 mg/kg/d

*** How to digitilgion ??**

8 hr 8 hr 12 hr
1/2 TDD → 1/4 TDD → 1/4TDD → M.D

*** Monitoring For Digitalis toxicity by :**

- ECG SB , HB
- Serum Digoxin level 0.8 – 2 ng/ml

*** Signs of Toxicity :**

A , N , V , diarrhea , weakness – visual disturbance – ECGHB , SB

TTT

- Stop drug
- Atropine for SB , HB
- For dysrrhythmia → Lidocaine
- Artificial Respiration .

B. other inotropic Drugs :-

Rapid acting, short duration catecholamine.

In sever CHF & RE dysfunction

Drug	Mg / kg / min	↓ S.E
Adrenaline	0.5	BP , A
Isoproteronolol	0.5	Pul , V.D
Dopamine	10	↑ HR ,A
Dobutamine	10	// //

(3) Vasa Dilators

After load ↓ agents

Drugs	Mg/K g/min	IV	S.E
Hydralazine	1	//	↑ H.R.
NG	2	//	-----
Na nitroprusside	4	//	Hepatic dysf.

Captopril - Neonate : 0.5 mg / kg / dose → 4 / d

- Child : 1 mg / kg / d

N.B

- * Hydralagine : Arterodilator
- * NG : Venodilaror
- * Captopril : Both

<i>PS</i> <i>AS</i>	
Types	
Supra ventricular - ventricular - sub ventricular	
Haemodynami	
- AS → LVH - Degree of H. Depend on i stenosis - sever As → LV Dilation → shock	- PS → RVH - PS - Sever AS → pat .F.O → Critical PS
C/P	
<u>SYMPTOMS:</u> * Mild → MOD → Asyptomatic. * Sever → - Easy Fatigability - Syncope * Critical → CHF <u>SIGNS:</u> • Sys .thrill at URSB • Ejection click . URSB • S2 Paradoxical splitting • Sys. ejscion murmur at 2 Rics propagate to Apex , neck • AR murmur • Critical AS → CHF → no murmur	<u>SYMPTOMS :</u> * Mild → asymptomatic. * Mod → - Easy fatigability Dyspnea, - Sever H.F. * Critical → Tachypnea <u>SIGNS:</u> 1 - ULSB 2- ULSB 3- Wide 4 -USB → back * Hepatomegally
INV	
* ECG : IVH * X-ray : - Prominent Aortic Knob - Cardiomegally - If sever → pul . V. congition * Echo :	* ECG : RVH , RT Axis Deviation * X-ray : Prominent main PA segment If sever : lung aoligemia . * Echo :
TTT	
* Mild → follow up * Mod → ballon diltaiion * Sever → valve replacement	* Ballon valvoblasty * AB prophylaxis * No need to Activity

PDA ASD	
Types	
Communication () Aorta , pulmonary artery	1. Ostium " premium, Secondium"commonest" 2. IVC-ASD 3. SVC-ASD
Hemodynamic	
* BL. Shunt From Aorta → pul .artery. * Shunt depend on (ducts size & PVR , SVR) * If small : Pul. Artery P. → Normal * If large :↑ to systemic level	* ASD → blood shunt from LT → RT Side " RV e' ↑ compliance " → RT Side * VOL. Overload → ↑ Pul. Artery p.
C/ P	
<u>Symptoms :-</u> - Small : → Asymp - Large : → HF , growth Retardation.	<u>Symptoms :-</u> - Small : → - Asymptomatic. - Large : → - F. To thrive - Exercise intolerance - HF
<u>Signs :-</u> • Apical Pulse : → prominent • Pulse p. : → Bounding – wide • Thrill : → - Sys . or continuous - 2nd Lt I.C.S - radiate to it clavicle • AUS :- - Continuous Machinery murmur - in 2nd Lt I.C.S : → Lt clavicle - Fun MS. murmur : mid diast. rumbling mur.	<u>Signs :-</u> • Lt pericardial pulg • RVH • Hyperdynamic Apex • AUS :- - Small : silent - Large : S2 Wide Fix Split هام - Ejection Systolic murmur. - Fun TS murmur : Mid diast. Rumb. Mur.
Comp	
* ECG : LVH * X – ray : LVH , PUL , plethora * ECHO : D * CATH. : in Aypical Finding	* R * R..... * *
INV	
* <u>HF</u> * <u>IE</u> * <u>Pul</u> or sys . emboli	<u>as VSD But :-</u> * late : - RVE - Eisenmenger Syndrome * ↓ I.E. risk " slow shunt " * paradoxical Embolization
TTT	
* Tran-catheter closure	* Surgical Closure

Q. D.D OF Generalized edema ?

D.D	Renal edema		Kwashiorkor Nutritional edema	Cardiac edema	Hepatic edema	Allergic edema	Protein Loosing enteropathy
	Nephrotic	Nephritic					
Age	2-10 yrs	2-12 yrs	6m-3yrs	Any age	Any age	Any age	Young age
History	Systemic disease or sudden onset	Strept. Infection	↓ Protein intake	CHD. Or Rh. Fever	Hepatitis or chronic liver disease.	Exposure to allergens	Growth failure
Edema	Severe with : - Hydrothorax - Ascites (anasarca)	Mild with puffy eyelids	Generalized with no serous effusion	Dependent edema starts in L.L. with ascites.	Ascites, then edema L.L.	Eyelids, lips, neck, Scrotum.	Dependent Generalized
Other finding	1. Albumin. 2. Cholesterol. 3. Proteinuria.	1. Red urine. 2. ↑Bp. 3. ↑ASO.	1. Mental changes 2. Muscle wasting. 3. Growth retardation.	1. Symptoms& signs of CHF. 2. ECG changes. 3. Gallop. 4. Neck veins. 5. Enlarged tender liver.	3- Shrunken Liver. 4- Jaundice 5- Palmer erythema 6- Spider naevi 7- Flabby tremors..	1- Itching. 2- Skin rash. 3- By anti - allergic.	↑Protein in stool.

Q. D.D of puffy eyelids:

- 1- As part of generalized edema (.....).
- 2- Local causes:
 - Conjunctivitis
 - Orbital cellulitis
- Chronic cough (B.A)
- Whooping cough
- Excessive prone sleep
- Hypothyroidism
- Cavernous sinus thrombosis
- Surgical emphysema.

AGN

Causes:

1. Infection :

o **Post-infectious GN:**

- o Post-strept. Infection MCQ the most common.
- o Following other infect (staph ,viral).

o **HIV associated nephropathy.**

2 G.N.:

- o Membrano-proliferative G.N.
- o Acute on top of chronic G.N.

3 auto-immune:

- o Anaphylactoid purpura.
- o SLE.
- o Good-Pasteur disease .

1_ رايح جاي

• **Recurrent gross hematuria syndrome :**

- o Idiopathic benign familial hematuria .
- o Hereditary nephritis.
- o Alport syndrome.

A-PS-GN

Acute post-streptococcal Glomerulonephritis

Def: Acute , specific , self limited , non-suppurative inflam. Disorder affect. Kidney, 1ry glomeruli due to immunological response to infection .

A/E : nephrogenic strains of group A-B hemolytic strept.

Ag-Ab reaction due to previous infection of skin or throat by nephrogenic strains of

	Pharyngeal infection	Skin infection
<i>Latent period</i>	(1 – 2) week	(2-3) week
<i>Serum Ab</i>	* Good Ab response * ↑ ASO	* Poor Ab response d.t skin lipid interfere e` streptolysin O antigenicity * ↑ Anti- DNase.

§ Evidence of immune mechanism:

- 1- Latent period () strept. inf. & onset of disease.
- 2- Serum complement .
- 3- detection of immune reactants.

Pathogenesis: Immune complex mediated

1- Strept. Infection:

Ag-Ab reaction → Ag /Ab complex

- Deposited in glomer. BM → Ag/Ab complex fixes complement .
- Production of chemotactic factors & inflammatory Process.

2-Both:

(deposits on BM +Prolifer.& swelling of endothelial cells of capillaries) →
Narrowing of capillary cavity → renal Ischemia → secretion of rennin →
angiotensin II → generalized VC.

3-Angiotensin : ++ aldosterone release → salt water retention.

C/P:**1) History & latent period:** Age (2-12yrs)

- Of streptococcal infection (sore throat ,scarlet fever, impetigo).
- After sore throat → 1-2 weeks.
- After skin infect → 2-3 weeks.
- Not > 4weeks → doubtful diagnosis.
- Not < 4 days → uncommon & indicate :
 - o Acute on top of chronic G.N.
 - o Recurrent gross Hematuria syndrome .

2) Constitutional symptoms: F H M A + pallor (anaemia).

3) Major symptoms: H₂O₂

o Hematuria :

- > 10 RBC`s HPF.
- The 1st & most common presentation (MCQ).
- Colour of urine : coca cola colour or smoky urine (due to prsence of RBC`s & formation of acid hematin).

o Hypertension:

- Bl.p.:
 - 1- Diastolic Bl.p.> 90 mmHg (< 10y) or >100 mmHg (>10y)
 - 2- Bl.p > 95% percentile for age .
- Sudden↑: in Blood P.may occure→ regular monitoring of Bl.p / 4hr.
- Usually: returns to normal at the end of 1st week ,or biphasic.

*** Causes:**

- 1- ↑ renin: generalized vasospasm.
- 2- ↑ Aldosterone: salt & water retention.
- 3- ↓ GFR: Hypervolemia.

o **Oliguria**: < 400 ml/day or even anuria (< 100 ml/d).

o **Oedema**:

§ Begins by puffiness in eyelids in morning , then oedema L.L. in night.

§ Minimal or moderate never severe (or never generalized) except with complications :

1 - Development of N.S.

1- Hypertensive HF.

2- ↑ Fluid intake with severe oliguria

§ Causes

1- Capillary permeability → d.to anoxia.

2- aldosterone → salt & water retention.

3- Hypertensive H.F.

4) Complications:

1- **Hypertensive Complications:** the most common complic. Of AGN. (MCQ).

- Hypertensive encephalopathy.
- Hypertensive H.F.
- Hypertensive retinopathy.

2- **Cardiac complications:** H.F. with enlarged tender liver or pulm. Oedema .

• Causes :

- hypertention : d.to. vasospasm.
- hypervolemia : salt & H₂O retention
- hyperkalemia : d.t. renal failure .
- Coronary spasm → Myocardial ischemia .

3- Cerebral complications : due to HTN.

	<u>Cerebral Ischemia</u>	<u>HTNsive Encephalopathy</u>
<u>Causes</u>	Sudden ↑ of Bl.P → vasospasm → Hypoxia .	Brain anoxia
<u>C/P</u>	Headache , vomiting , drowsiness , convulsions , diminution of vision.	- Irritability, Convulsions , vomiting , Headache . - The easiest compl. to be prevented & ttt by monitoring.

4- Renal complications:

- Chronic GN (5%)

- ARF(5%)

*Investigations :*1- Urine analysis:v Macoscopic :

- Volume: oliguria (< 400 ml / d).
- Aspect & colour: hematuria : smoky or coca cola urine.
- Specific gravity: ↑↑(1035).
- Protein: mild proteinuria (< 1 gm/d) → frothy.

v Microscopic:

- Cells:
 ý RBC`s: > 10 HPF (characteristic) & distorted in shape.
 ý WBC`s: ↑↑ in number.
- Casts: red casts: diagnostic for glomerular hematuria.
- Addis count: more accurate determination of No of RBC`s & WBC`s & casts.

كلمتان خفيفتان على اللسان ثقيلتان في الميزان حبيبتان إلى الرحمن

﴿ سبحان الله وبحمده سبحان الله العظيم ﴾

2- Blood analysis:

- Mild normocytic normochromic anaemia due to :

- 1- ↓ Renal erythropoietin.
- 2- Hematuria .
- 3- Hemodilution.
- 4- Autoimmune hemolytic anemia.

§ ↓↓ Complement (C_3 , C_4) characteristic : return to normal (3- 4) weeks.

3- Renal faunction test:

- Glomeular F.T.: ↑ creatinine & urea concentration.(impaired).
- Tubular F.T.: normal.

4- Renal imaging.: U/S**5- Renal biopsy:**indicated in atypical presentation - resolution.

- 1- abesence of evidence of strept. infection .
- 2- persistence of marked hematuria, proteinuria or both.
- 3- Normal complement.
- 4- Persistence of low C_3 > 3 months.
- 5- Development of ARF or chronic GN.

6- Investig .for the cause:

(evidence of strept. Infection) as Rh. Fever →

Anti-DNAse B: the best single test

- (exclude other causes of hematuria)
 - ý SLE: ANA (anti-nuclear Ab)→ Alport syndrome → test mother urine for blood & hearing test
 - ý Stones : U/S

D.D + causes of Red Urine

N.S. البيجيلة H A S H E C H اللي بيشر ب حشيش

H	Haemolytic uraemic syndrome	- The most common causes of R.F. in children. - E-coli infection → hemolysis → uraemia.
A	Acute Pyelonephritis	- Fever , toxaemia , loin pain. - No HTN. - Pus & bacteria in urine.
S	Sulfonamide Crystalluria.	- Symptoms disappear when stop the drug.
H	Hereditary nephritis (alport synd).	- Familial ,benign. - Hearing test , test the mother urine for bl.
E	Embolic Nephritis	- Complication of Inf. Endocarditis . - +ve blood culture . - ttt given → - ve bl. Culture & hematuria disappear.
C	Collagen disease (SLE)(lupus nephritis).	- Anti Nucleous Ab
H	Henoch – Schoenlein purpura (anaphylactoid P.)	- Purpura . - Joint manifestation. - Abd.manifest.: nausea,vomiting , abd.pain. - <u>Normal</u> serum C3

لا تمشي في طريق من طرق الحياة الا ومعك سوط عزيمتك وإرادتك لتلهب به كل
عقبة تتعرض لطريقك نيتشه

Clinically**Laboratory**

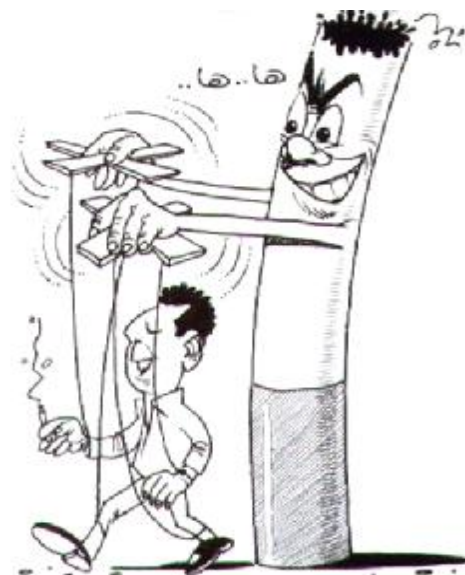
	AGN	NS
1- strept. Infect.	+ ve	- ve
2- Hematuria.	++	rare , transient
3- HTN.	++	rare , transient
4- Oliguria .	marked.	during oedema.
5- Oedema.	Minimal	Massive
1- S . Albumin.	Normal	Low
2- S.Cholesterol.	Normal	High
3- S.C3 &C4.	Low	Normal
4- Porteinuria.	Minimal	Heavy
5- Casts.	Red & granular	Hyaline
6- ASO .	Usually + ve	- ve
7- Renal funct.	Impaired	Usually normal

Course & Prognosis:

- § 90:95% → Complete recovery
- § 5%→ pass to subacute or chronic GN.
- § 5%→ Death d.to :
 - ý R.F.
 - ý H.F.
 - ý HTNsive encephalopathy.

TTT:1- Hospitalization & Bed rest:

- § Hospitalization is essential if complicated for monitoring.
- § Bed rest is avoided only during acute illness till :ptn free from oedema (for 2-3 weeks). → it is good for prevention of H.F. , HTN.



2- Diet:§ Fluid & salt:

ý Restricted in (oliguria – marked oedema –CHF).

ý Amount of fluid (400 ml/m²/d + amount of urine passed in every day).

§ Protein:

ý Restricted in (oliguria – anuria – R.F.) Only 0.5 gm / kg /day.

3- Drugs: (Antibiotic therapy):-

ý Procaine penicillin 6000.000 u/d × 10 d to eradicate strept.inf.

ý No maintenance dose as no recurrence.

4- Observation :

* B.P every 4 hours for fear of sudden ↑↑. * Urine output.

* Body weight daily → for oedema .

5- ttt of complications: من الجدول القادم6- follow up: 4w→ 6m * 4m→1yr. * 1yr→ afterwards.

HTN	H.F	ARF.
↓ < 140 / 90 > 140/90 ↓ ↓ ↓ No ttt e`out      e` Only E n c e p h a l o p a t h y Follow ↓ ↓ Up <u>Diuretics</u> <u>CCB</u> Lasix 2mg/kg/d Nifedipin ↓ Of Choise ↓ + If no response Anti-convulsant Add ACEI phenobarbitone	<u>ttt of HTN</u> Restrict salt & fluid. Diuretic: lasix I.V. Digitalis: (Rapidly) Dialysis (peritoneal) in refractory cases. Dilators.	1- <u>Diet:</u> restrict fluid& salt & protein. 2- <u>TTT of hyperkalemia:</u> - <u>Orally or retention enema:</u> Kation exchange resin. - <u>I.V.:</u> * Na HCO₃. * Ca gluconate. * Insulin + 50% glucose . If persistent → dialysis 2- <u>Correction of metabolic acidosis:</u> NaHco₃ 3- <u>Peritoneal dialysis</u> In severe cases.

Nephrotic Syndrome (N.O.)

Def : Clinical syndrome characterized by :

- 1- Generalized oedema.
- 2- Heavy proteinuria $>2\text{gm/d}$.
- 3- Hypoproteinaemia $< 5 \text{ gm/dl}$.
- 4- Hyper lipidaemia $>300 \text{ mg/dl}$.

Pathphysiology :

- 1- **Proteinuria** (1ry abnormality) $> 2\text{gm/d}$
Causes : $\uparrow$ Glomerular capillary permeability By unknown cause , or
Loss -ve charged glycoprotein. , or $\uparrow$ pore size of B.M.
Either : selective or non- selective.
- 2- **Hypoproteinaemia** ($< 5 \text{ gm/dl}$) $\rightarrow$ mainly hypoalbuminaemia($< 2.5 \text{ gm/dl}$)
Cause: loss of protein in urine.
- 3- **Generalized oedema**: causes :
 - 1- Hypoalbuminaemia .
 - 2- $\uparrow$ ADH $\rightarrow$ water retention.
 - 3- Activation of Renin –Angiotensin- Aldosterone system $\rightarrow$ Na & water retention.
- 4- **Hyperlipidaemia** : Causes: $\uparrow$ all serum lipids by :
 - 1- $\uparrow$ lipoprotein formation in liver due to hypoproteinaemia.
 - 2- $\downarrow$ lipid catabolism as lipase enzyme lost in urine .
 - 3- Hypothyroidism
- 5- **Hypocalcaemia**:
Causes: * $\downarrow$ plasma protein .
* $\downarrow$ GIT absorption due to edema.

6- Hypokalemia

Causes :

- 2^{ry} hyperaldosteronism
- ↓ intake by anorexia .
- ↑ loss due to diuretic therapy.

With Best Wishes
 د/ ماذح & د/ الجيزي

A/E , Classification of N.S. :

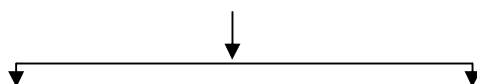
- 1st 3 months: congenital (finnish type).
- 3-12 months: Infantile type.

1- 1^{ry} Nephrotic s.(idiopathic ,lipoid) : 90 %**- Types acc. to steroid:**

- steroid responsive.
- Steroid non- responsive

- Types Acc. to histopathology:

- MCN.S. (85 %) : best prognosis.
- Diffuse mesangial proliferation (5%).
- Focal glomerular sclerosis (10%).

2- 2ndry N.S: 10% due to**Renal diseases**

- 1- Chr. Glomerulonephritis.
- 2- Chr. Pyelonephritis
- 3- Drugs: penicillamine , NSAIDS

Systemic diseases

- 1- Blood: A.pupura , S.C.anaemia.
- 2- Infection: T.B, Bilhaziasis .
- 3- Metabolic: D.M., amyloidosis.
- 4- Allergic: serum sickness.

Histological types:

- * Membranous. * Focal sclerosing type + evidence of the cause :
- * Proliferative. - DM → Microangiopathy
- * Membrano- proliferative. - Amyloidosis

Primary N.S (idiopathic)

A/E : Unknown.

C/P : 1- Onset → insidious.

2-Constitutional symptoms →

- Fever , headache , malaise.
- Anorexia, nausea , vomiting , diarrhea
- Pallor due to oedema.
- Hepatomegaly due to oedema & fatty infiltration of liver cells.

3- Oedema (main presentation)

- Characteres → Generalized , soft , pitting , seen on shin of tibia & Overlying skin: tense , shiny & stria of abd. Wall.
- Course
 - § 1st morning edema of eyelids → bloated face.
 - § Ankle edema → generalized edema in ext. genitalia , neck , limbs , Ant. Abd. Wall.
 - § In severe case it involve:
 - ý Thigh → pressure by cone of stethoscope to skin (pitting).
 - ý Ant.Abd. wall → prominent underclothes marks
 - ý Scrotal oedema in males.

4-Urinary changes.

- a) Haematuria : 5% of cases .
- b) Oliguria : due to bed rest or stress or steroids
Urine Output $\propto$ 1/ degree of oedema.
- c) proteinuria : leads to frothy urine .

5- Others

- § Hypertension : 5-10%.
- § Azotaemia : absent or minimal.

§ Hypocalcemia.

§ Hypokalemia.

6-Respiratory difficulty due to

§ Massive ascites.

§ Pleural effusion.

§ Chest infections.

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Investigation:

1- Urine analysis

§ Volume : oliguria.

§ Aspect : frothy

§ S.G.: ↑↑.

§ Proteinuria: moderate or massive selective in MCNS mainly albuminuria.

§ Casts: hyaline , granular , lipoid.

§ Haematuria microscopic haematuria.

§ Urine culture.

2- Blood exam : for

1- Hypoproteinaemia

§ Total level < 5 gm/dl.

§ Inversion of A/G ratio.

2- Hypercholesterolaemia : > 300 mg/dl.

3- Electrolytes.

* ↓ Na : dilutional hyponatraemia.

* ↓ K d.to ↑aldosterone , diuretics , ↓ intake .

* ↓ Ca d. to loss of Ca bound albumin

4- CBP

§ RBCs→ ↑ Hb & HCV but no anemia .

§ TLC → normal.

§ Platelet count normal but ↑aggregation .

5- C3 level : normal

6- Serum triglycerides & ESR : ↑↑

3- **Renal function tests:**

normal in idiopathic type Unless patient develop renal failure .

4- **Renal biopsy :**

Indication : سؤال هام شفوي ونظري

1- Abnormal age : < 2 yrs Or > 10 yrs.

2- Abnormal presentation → haematuria Or Azotaemia.

3- Abnormal Lab. Finding.

§ ↓C3 .

§ proteinuria persistent > 1 month.

4- steroid dependent & resistant cases.

5- Frequent relapse .

Complications : A B C + S / R

§ 5 A

▼ Acute R. failure (esp.FGS)

▼ Anaemia (loss of transferrin in urine).

▼ Atherosclerosis.

▼ Abdominal pain .

▼ Abnormal growth & Nutrition.

§ 1 B Bleeding tendency (loss of clot. Factors 9, 11 , 12).

§ 2 C

▼ Coagulation abnormality & thrombotic tendency .

ý ↓ antithrombin III.

ý ↑ platelet aggregation.

ý ↑ inhibitors of fibrinolysis.

✓ Cataract (d.to steroids).

§ 2 S

✓ Shock (Hypovolemic d. to rapid diuresis).

✓ Skin infection & diseases .

§ 1 I → ↑ infection liability (the most common C/O).

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✓ D.to :

ý Immuno-suppressive drugs.

ý ↓ bactericidal activity of WBC`s .

ý defective oppsonization.

ý Hyposplenism.

✓ the most common infection (pneumococcal peritonitis) .

✓ the most common site (peritoneum , skin).

✓ the most common organism (pneumococci , E-coli).

§ 2 R

✓ Relapse (either lab. → heavy proteinuria & or clinical → oedema) .

✓ Renal – tubular disorder.

TTT:

1- **Prophylactic:** if 2ry (ttt of DM.)

2- **Curative:**

A- Specific ttt

1. **Bed rest:** Contraindicated:

§ To avoid thrombosis.

§ To mobilize oedema fluid

1- **Diet:**

§ **Fluids :**

ý Give amount of urine out put .

ý + give amount of vomiting , diarrhea.

ý + 500 cc for insensitive loss.

ý + 500 cc for each↑ in temp. 1°C.

§ Salt: restricted .

§ Protein: ↑↑ + salt-free albumin .

§ CHO: freely advised.

§ Fat : better avoided .

2- Drugs:

1- Antibiotics :

§ Prophylactic: against inf. by pneumococci.

§ Therapeutic: for ttt of any inf. till result of cult. & sens . Test

2- Vaccination:

§ Polyvalent Pneumoccal Vaccine.

§ H. influenza vaccine.

3- Anti coagulant:

§ D.to hypercoagulability .

§ Avoid heparin (loss of anti-thrombin III).

4- Diuretics:

§ Indications :

ý Start of the disease till steroid induced diuresis.

ý Steroid resistance.

ý Sever oedema with ascites or pl.effusion.

§ Types:

ý Mild to moderate oedema: clorothiazide +_spironolactone.

ý Sever oedema : furosemide + spironolactone + I.V. salt free albumin.

5- Steroids : In :

ý 1 ry N.S.

ý If collagen disease.

6- Immuno suppressive drugs:

§ Drugs: azathioprine , cyclophosphamide.

§ Indications:

ý Steroid resistant .

ý Steroid dependent.

ý Frequent relapse with contraind. steroids.

§ S/E:

ý Alopecia

ý B.M. depression.

ý Sterility.

ý Varicella infection.

§ Duration: 6-12 weeks

7- TTT of special clinical situations:

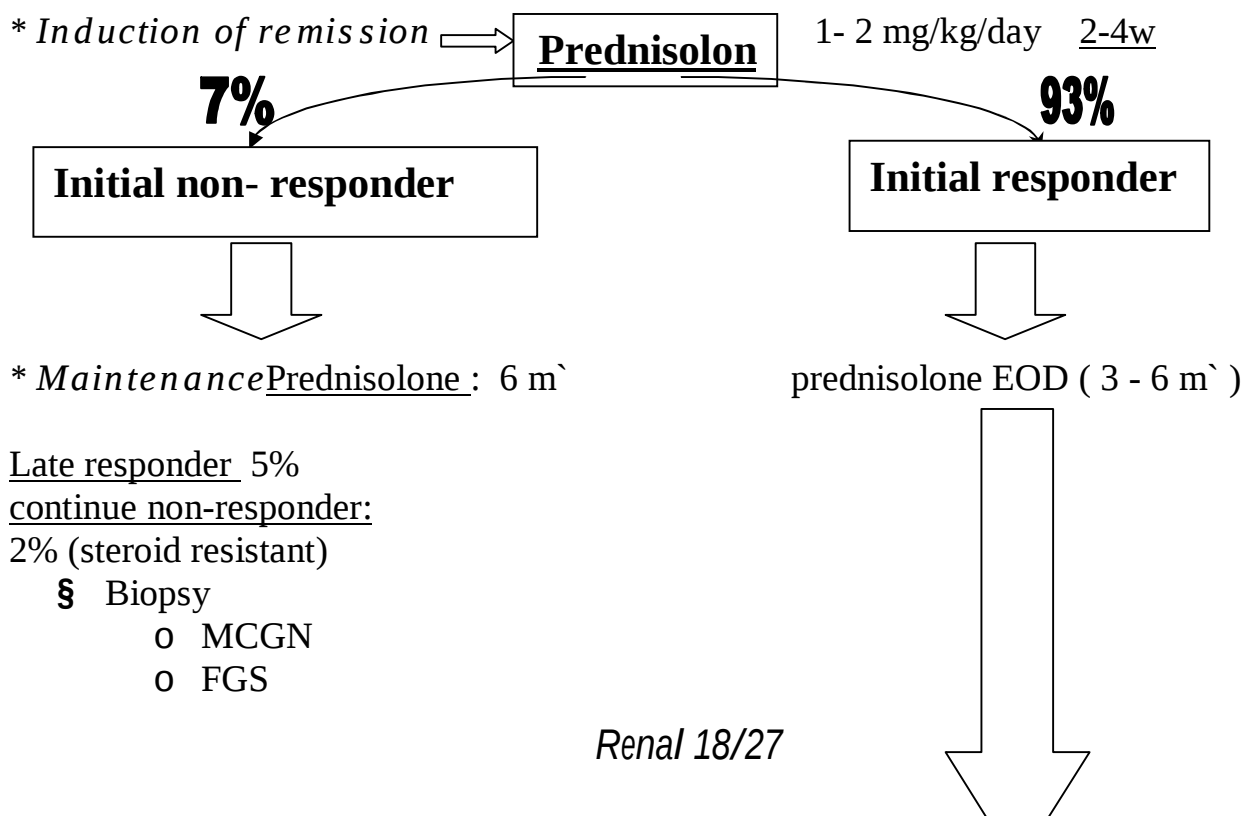
1- Infrequent relapse: induction & maintenance.

2- Frequent relapse & steroid dependent: Imm.sup. drugs .

3- Steroid resistance:

§ MCGN → steroid + Immunosup. (6 – 12 w).

§ FGS → prednisolone+ vincristine + cyclosporine.



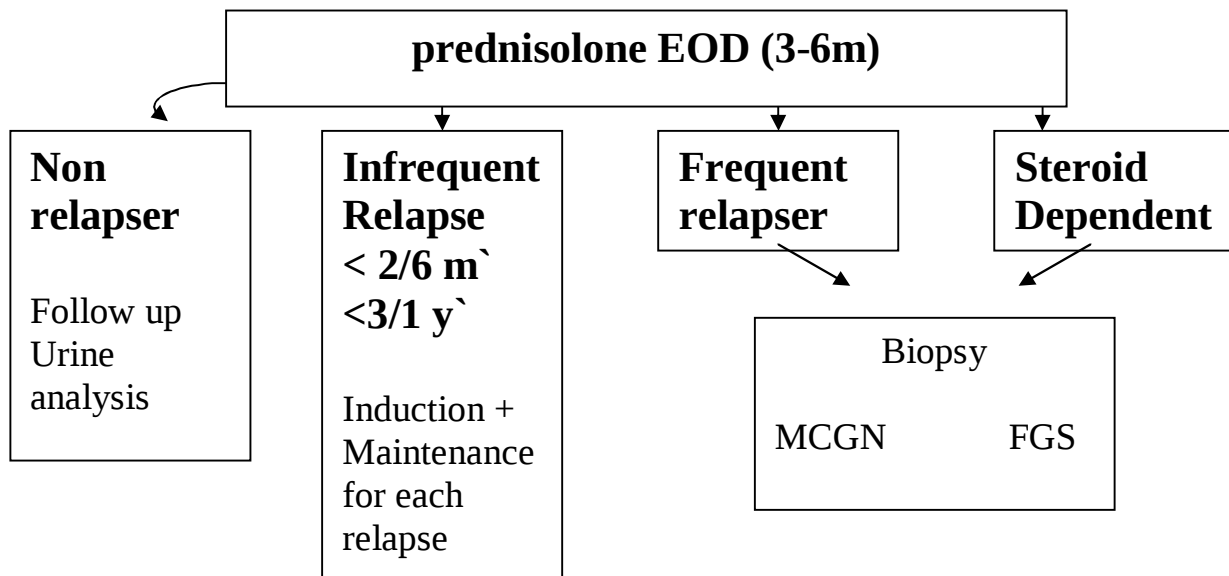
ý Late responder 5%

ý continue non-responder:
2% (steroid resistant)

§ Biopsy

o MCGN

o FGS



B- Surgical ttt: Renal transplantation.

U.T.I

A/E:

1- Causative organism: normal rectal & perineal flora:

§ Colonic bacteria:-

- ▼ E-coli (80 %)
- ▼ Proteus
- ▼ Klebsiella

§ Normal skin inhabitants:

- ▼ Staph. Epidermidis.
- ▼ Staph. saprophyticus.

§ Others:

- ▼ Bacteria: Salmonella.
- ▼ Viral

▼ Fungal.

▼ Parasitic: schistosomiasis.

2- Route of infection:

§ Neonatal period : Blood stream , urethra.

§ Later in life: trans-urethral.(ascending)

3- Predisposing factors: O₂ C U P

§ Obstruction of U.T.

§ Oxyuriasis.

§ Constipation.

§ Urethral instrumentation.

§ Poor hygiene.

C/P:

1- Asymptomatic : accidentally during routine analysis & culture.

2- Symptomatic:

a. Non-specific manifestations: F H M A + N V D common in infancy.

b. PUO: persistent > 2weeks.

c. Classic presentation:

§ Disturbance of act of micturation (urgency , frequency , dripping ,incontinence).

§ Foul smell of urine.

§ Hematuria.

§ Abd. Pain.

§ Manif. Of infection.

§ Complications.

3- C/O:

§ HTN → d.to renal scarring.

§ CRF → d.to renal damage.

من أذكار الصباح والمساء :

سبحان الله وبحمده عدد خلقه ورضا

نفسه وزنة عرشه ومداد كلماته ﴿

لو وزنت بما قلت منذ اليوم لوزنتهن .

رواه مسلم

Investigations:

TTT:

1- Prophylactic:

- § Good hygiene to avoid ppt.
- § ↑ Water & fluid intake.
- § Frequent emptying of bladder.
- § Change urine PH.

2- Curative:

- § Supportive (ARSEF)
- § Symptomatic.....
- § Specific measures.....

1. Antibiotics:

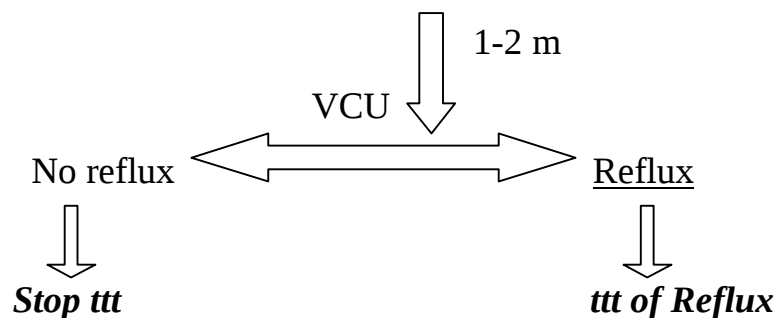
Acc. To culture & sens. Test → (7 – 10 days).

- § Ampicillin (100mg/kg/d)
- § Amoxicillin (50 mg/kg/d)
- § Septazol
- § Ciprofloxacin (3rd gen. Quinolone) the best

2. Chemoprophylaxis:

After c/p improvement → stop A.B. 1/w →  repeat culture

Chemoprophylaxis (1/3 dose of therapy) repeat ttt acc.to New cult. & sens.



* Grade I,II,III:- Prolonged prophylaxis + regular follow up by VCU till it disappears.

* Grade IV,V:- surgical ttt.

3- TTT of complications:-....4- Follow up:-

§ Urine culture at 3 months interval for 1-2 yrs even if child asymptomatic.

§ Recurrent attacks of UTI with U.T. obstr. → prolonged prophylaxis + surgical ttt .

5- Management of VUR.

Causes of Hematuria

HAT HAT + U (UTI) S (Stone) A (AGN) تتلخص في

Hemorrhagic Blood DES

Angiomatous Malformation

Trauma

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Hemorrhagic Fever (HTN)

Aneurysm rupture

Tumor

Causes of Red urine

- 1- Hematuria.
- 2- HBuria.
- 3- Hepatitis
- 4- Hepatitis (jaundice → bile).
- 5- Porphyrin.
- 6- Food
- 7- Food coloring material.
- 8- Drugs : Rifampici

	<i>Lipoid Nephrosis</i> (1 ry nephrotic syndrome)	2ry N.S. (nephritic - nephritic synd)
Age	2- 10 yrs	Usually > 5yrs
HTN Hematuria Azotemia	Absent or Minimal	Present
Cast	Hyaline	RBC`s + Hyaline
Proteinuria	Selective albumin	Non-selective (alb. – glob.)
C3	Normal	↓↓↓
Steroids	Sensitive	Dependent or resistant
Prognosis	Good	Guarded

Approach to Diagnose case of Proteinuria

Def : Urine of child has protein (40 % Albumin & 40 % TH Or. 15% Igs , 5% Others).

A- Confirmation of presence of proteinuria:

By detection of protein in urine by:

- 1- Dipstick: detect albumin only .
- 2- Sulphosalicylic acid: detect all proteins.
- 3- Protein / Creatinine ratio.
- 4- Quantitative method.

ý Nephrotic: > 40 mg /m²/ hour.

ý Non- Nephrotic: 4 - 40 mg /m²/ hour.

B- Exclusion of other causes of false + ve tests of proteinuria:

- 1- Highly concentrated urine .
- 2- Alkaline urine .
- 3- Pyuria .
- 4- Gross hematuria .
- 5- Skin cleanser . (Chlorhexidine).

C- Differentiate () types of proteinuria:-

	<u>Glomerular</u>	<u>Tubular</u>
1- Dipstick	+ ve	- ve
2- Quantity	Mild	Severe
3- Protein	Mainly albumin	Low M.Wt. protein.
4- Tubular Function	Normal	Abnormal

D- Causes of proteinuria:

- 1- Transient : exercise , fever , H.F. , anemia .
- 2- Orthostatic : common in young female(10-12 yrs)
- 3- Persistent :
 - § Over flow: myoglobin , light chains .
 - § Tubular .
 - § Glomerular:
 - ý Congenital
 - ý Acquired
 - ü 1ry :idiopathic .
 - ü 2ry : drug , Infection

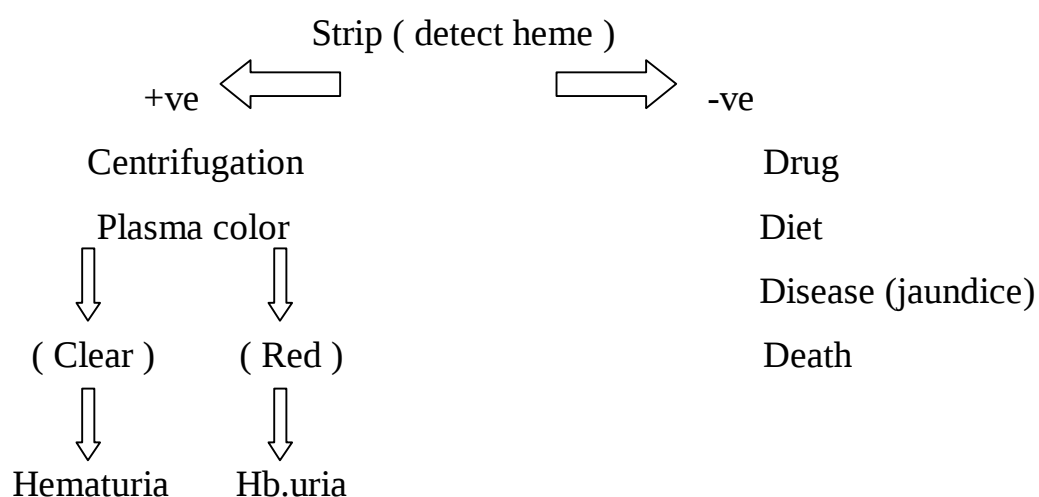
Approach D Of A case Of Hematuria

Def: presence of abnormal RBC's in urine.

Detection :

- 1- **Gross hematuria:** red urine
- 2- **Dipstick:** change color of strip.
- 3- **Microscopically:** RBC's in 10 ml urine ≥ 5 / HPF.
- 4- **Addis count:** > 240.000 RBC's in 12 hrs. urine.

Work up of red urine:



Types of Hematuria :

	<i>Non –glomerular</i>	<i>Glomerular</i>
<i>Colour</i>	Red , bright	Smoky , coca cola
<i>Clot</i>	Present	Absent
<i>Cast</i>	Absent	Present
<i>Proteinuria</i>	Absent	Present
<i>RBC's morphology</i>	Normal	Dysmorphic

Dr.Madeh Dr.AlgizY

Q. Factors affecting Growth ??

1- Intrauterine:

a-: 1st trimester : organogenesis

- Chromosomal abrasion
- Irradiation
- Drugs: teratogenic
- Infection: Toxoplasmosis , Rubella , CMV.

Hazards:

- abortion
- anatomical abnormality

b- fetal period (2nd or 3rd trimesters):-

- 2nd trimester : $\uparrow$ in length .
- 3rd trimester : $\uparrow$ in wt .
 - o Mother health or Nutrition.
 - o Dse of cord or placenta .
 - o Dse of uterus or amniotic sac .
- Hazards:
 - o Still birth .
 - o Premature delivery

2- Extra – uterine :

a-Neonatal period: (1st 4 weeks of life)

- * diseases :
- 1- Birth trauma.
 - 2- Blood group incompatibility.
 - 3- Congenital anomalies.
 - 4- Failure to initiate respiration
 - 5- Infection.

- b- Infancy period:
- Early : 4 m` → 1 year
 - Late : 1 yr → 2nd year

• **Criteria :**

- o Weaning (malnutrition. . .).
- o ↑ Rate of growth.
- o Eruption of all milk teeth.

c- Childhood period: social & educational maturation

- d- Adolescence:
- Puberty.
 - 2ry sex characters.
 - Growth spurts.

Q. Assessment of growth parameters ?

1 - Wt	• <u>At birth</u> : 3 – 3.5 kg • <u>3 -12 m:</u> = age + 9 / 2 kg. • <u>> 12 m:</u> = (age (yrs) × 2) + 8 kg
2- Length	• <u>At birth</u> : 50 cm - <u>at 1 yr</u> : 75 cm • <u>At 1st 3m:</u> 3 cm / m` - <u>at 2 yrs</u> : 87 cm</li> <li>• <u>At 2nd 3m:</u> 2 cm / m` - <u>at 4 yrs</u> : 100 cm • <u>At 7-12m:</u> 15 cm / m` - <u>2 yr`</u> = (age × 5) + 80 cm
3-Skull circumf.	• <u>At birth</u> : 35 cm • <u>At 6m</u> : 42 cm • <u>At 1yr</u> : 45 - 46 cm • <u>At 2yr</u> : 49 cm • <u>At 3yr</u> : 50 cm
4- Fontanel	<u>Post. Fontanel</u> : closed at birth ,or soon after 2m. <u>Ant. Fontanel</u> : - <u>at birth</u> : 4-5 cm - <u>at 1yr</u> : 1.5 cm - <u>at 6 m</u> : 3 cm - <u>at 1.5 y`</u> : closed

5- Teeth	<u>Milk Teeth (20)</u> 6 m → lower central incisors 7 m → upper central incisors 8 m → upper lateral incisors 9 m → lower lateral incisors 12 m → 1 st molar 13 -18m → canines 24 -30m → 2 nd molars	<u>Permanent teeth (32)</u> 6 yrs → 1 st molars 7 yrs → central Incisors 8 yrs → lateral Incisors 9 yrs → canines 10 yrs → 1 st premolars 11 yrs → 2 nd premolars 12 yrs → 2 nd molars 17 yrs → 3 rd molars (wisdom)
6 - MAC	• <u>Normally</u> : → > 13.5 cm. • <u>Borderline undernutrition</u> → 11 - 13.5 cm . • <u>Undernutrition:</u> < 11 cm	
7- Chest circum	• <u>At birth</u> : Chest Circumference < Head Circumference • <u>At 6m:</u> > 1	
8 - chest cir./ Head circumferenc	• <u>At birth</u> : < 1 • <u>At 6 m</u> : = 1 • <u>After 6m</u> : > 1	
9 -Triceps skin fold	• <u>At birth</u> : 7 mm • <u>At 1 yr</u> : 12 mm	
10 - body ratio US/LS	• <u>At birth</u> :1.7 • <u>At 3yrs</u> : 1.3 • <u>At 9 yrs</u> : 1	
11- Osseous Growth	• No. of carpal centers = age (yrs) +1 • 1st 6m : 1st carpal centre . • 2nd 6m : 2nd carpal centre .	

Q. Developmental milestones ?

Age	Gross motor	Fine motor vision	Language hearing	Social emotional
At birth	Flexion on prone	Blinking to flash light	Responds to bell	
1 m	Raise Head slight on prone	* follow moving Object. * Respond to sounds by Blinking.	Vocalizes	Looks to faces
2m		Follow 180 by eye		- Recognize mother. - Social smile.
3m	Head support	Regards his hands	Pronounce coo	Anticipate feeding
4m	- sit with support. - Moro disappear	Grasp rattle	Laughs loudly	Enjoy looking around
6m	- Sit without support . - start crawling	- grasp by palm. - Reach objects & bring to mouth		A ware from strange faces
9m	- Creeping. - Stand e` support	Grasp by thumb & fingers.	Say dada & mama non - specific	Responds to his name
12m	- Stand freely - Start walking e` one hand held	Hold cup for drinking	- say dada, mama non - specific. - 1-3 meaningful words	Play ball e` the examiner

15m	- Walks freely . - crawls upstairs	- draw a line . - build a tower of 2 cups	Baby own language(jargon)	Imitates house work
18m	- start running - walk upstairs e on hand held.	- build tower of 3 cubes. - Imitate vertical strokes	Average 10 words	Play e other children
2yrs	- runs well. - Walk upstairs one step at once.	- build tower of 6 cubes. - Imitate horizon strokes.	Join 2-3 words together.	Know his family members & their names.



دعاء " سيد الاستغفار " :

اللهم انت ربى لا اله الا انت خلقتنى وانا عبدك وانا على عهدك ووعدك ما استطعت اعوذ بك من شر ما صنعت ابوء لك بنعمتك على وأبوء بذنبي فاغفر لى فإنه لا يغفر الذنوب إلا أنت .

(من قالها من النهار موقنا بها فمات من يومه قبل ان يمسى فهو من اهل الجنة ومن قالها من الليل وهو موقن بها فمات قبل ان يصبح فهو من اهل الجنة)
... حديث شريف رواه البخارى فى صحيحه

Mode of inheritance

Single gene			- Mendelian inheritance		- 2 Allele identical à Homozygous
					- 2 Allele not identical à Heterozygous
Autosomal هَام			Sex linked هَام		
	Dominant	Recessive	A)Y-chromosome		
1- gene show It`s Effect in	Homozygous Heterozygous	Homozygous only	Trait appears <i>only in male</i> . Transmitted from father to <i>All Male</i> offsprings.		
2 -Affected person parents	Affected	Not Affected	B) X- Chromosome inheritance		
3 -Each Offspring of	1 aff.. Parent has <u>50%</u> chance to be affected	Carrier person has <u>25%</u> chance to be aff.	1- dominant	2- Recessive	
	Unaff..Relatives will not have aff. offsprings	+ve consanguinity	- <i>Affected F.</i> transmit. Affected gene to : 50 % sons ,50% daughters.	- Gene carried by <u>Males</u> , <u>Females</u> <u>But</u> i Aff. Person is <u>Males</u> .	
4 -eg	*Achondro_ Plasia . *Marfan syn.	*Thalassemia *Fanconi anemia *Lignac syn.	- <i>Affected M.</i> transmit Affected gene to all daughters non of his sons e.g: Hypophosph. Rickets. Alport Syndrome <u>3- sex limited</u> - gene expression in only one sex not i Other . e.g: gene Det. Heavy beard <u>4- sex controlled</u> - gene express itself in diff. manner in both sexes. e.g : gene determine Voice ♂ : low pitched ♀ : high pitched	- <i>Never</i> transmit from father to his sons . e.g.: - Hemophilia A.B - G6 PD - Ms Dystrophy	
3- Incomplete Dominant		4- Co-Dominant			
Gene partially expressed in Heterozygous state . e.g sickle cell Anemia . gene HbS :- Homozygous : S.C.Anemia Heterozygous : An. only in ↓ O2		*2 Allels (Dominant, Not identical) *Each show it`seffect . * e.g ABO group : AB group is Heterozygous AB			

Multifactorial
<u>Def</u> : inheritance
caused by many genes(polygenic) plus effect of environment
<u>Normal variation</u> :
i magnitude of trait determined by
- number of genes
- Number of environm. factors
<u>Examples:</u>
A- Normal trait :
Stature
Intelligence
Bl.P.
Skin colour
B- Abn. Trait
B.A
D.M
P.U
Coronay Ht. Dse
Schizophrenia

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Intelligence
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Skin colour

B- Abn. Trait

B.A

D.M

P.U

Coronay Ht. Dse

Schizophrenia

Mode of inheritance

Single gene			Multifactorial	
- Mendelian inheritance - 2 Allele identical → Homozygous - 2 Allele not identical → Heterozygous			Def: inheritance caused by many genes (polygenic) plus effect of environment Normal variation: i magnitude of trait determined by - number of genes - Number of environm. factors Examples A- Normal trait : Stature Intelligence B.L.P. Skin colour B- Abn. Trait B.A D.M P.U Coronay Ht Dse Schizophrenia	
Autosomal			Sex linked	
			A) Y-chromosome	
1- gene show It's Effect in			Trait appears <i>only in male</i> . Transmitted from father to <i>All</i> Male offsprings.	
			B) X- Chromosome inheritance	
			1- dominant	2- Recessive
2 -Affected person parents	Dominant Homozygous Heterozygous Affected	Recessive Homozygous only Not Affected	- <i>Affected F.</i> transmit. Affected gene to : 50 % sons ,50% daughters. - <i>Affected M.</i> transmit Affected gene to all daughters non of his sons e.g <i>Hypophosph. Rickets.</i> <i>Alport Syndrome</i> 3- sex limited - gene expression in only one sex not i Other . e.g: gene <i>Det. Heavy beard</i> 4- sex controlled - gene express itself in diff. manner in both sexes. e.g : gene determine Voice ♂ : low pitched ♀ : high pitched	
3 -Each Offspring of	1 aff.. Parent has 50% chance to be affected	Carrier person has 25% chance to be aff.	- Gene carried by <u>Males</u> , <u>Females</u> . <i>But</i> i Aff. Person is <u>Males</u> .	
4 -eg	* <i>Achondro _ Plasia</i> . * <i>Marfan syn.</i>	* <i>Thalassemia</i> * <i>Fanconi anemia</i> * <i>Lignac syn.</i>	- <i>Never</i> transmit from father to his sons . e.g.: - Hemophilia A.B - G6 PD - Ms Dystrophy	
3- Incomplete Dominant	Gene partially expressed in Heterozygous state . e.g sickle cell Anemia . gene HbS :- Homozygous : S.C.Anemia Heterozygous : Anemia only in ↓ O2 condition .	*2 Allels (Dominant, Not identical) *Each show it's effect. * e.g ABO group : AB group is Heterozygous AB		

Types of chromosomal abnormality

- 1- Numerical : * Autosomal Abn. à Trisomy 18 Edward 21 Down 13 Patau__ 8
 * Sex.ch. Abn. à Turner & Klinefilter
- 2- Structural : * Autosomal à 5 p Cri-Du-chat
 4p wolf ,13q , 18q , 21p
 * Sex ch. Abn à long arm detection of chr.x
 Short arm detection of chr.x
 Ring x chromosomal

Sex Ch. Abnormality

1- Turner syndrome بنت

- Incidence 1/ 250
- Karyotype XO
- C/P

short stature

Mental subnormal : (IQ may attain high levels)

Face : Hypertelorism – narrow maxilla – small mandible – low set ears

Neck : webbed

Nipples : wide spaced

Extremities : Cubitus valgus , Lymphoedema in the Hand and dorsum of foot .

Congenital anomalies :- 35% - Coarction of aorta

Congenital Renal Dse.

Infertility dt ovarian dysgenesis

2- Klinefilter (xxy) syndrome شاب عاقل

MCQ : most common X chromosomal Abnormality .

Incidence : 1 : 1000

Karyotype : 47 (xxy)

- C/P** :
- phenotype male e' gynecomastia
 - Mentality à Normal.
 - Small Testis
 - Sterility

Dysmorphology Disorders

Def : Abnormality of body structure Originate before birth .

Pathogenesis :

1- Malformation

Def Arrest of normal development of an organ

AE Intrinsic abnormal developmental process

E.g. ASD, VSD, cleft (palate , lip)

2- Deformation

Def Abn. (form – shape – site) of part of body w' differentiate normally.

AE Mechanical Factor

E.g. Craniostenosis – Hip Dislocation

3- Disruption

Def Morphological defect of (organ - Part of organ – region)

AE Extrinsic breakdown

Mech

1 - Aminotic band :

- Amnion rupture → Amniotic band Adhere to fetus on growth band comparison developing area → resorptive Necrosis
- e.g. i bands affect usually :

Limbs à Amputation of digits or arm .

Craniofacial à Encephalopathy.

2 - Vascular disruption :

AE : 1- Abnormal develop. of fetal Bl.V .

2- Placental circulation disruption.

3- Maternal ↓ Bp .

Result Early gestation à Atresia Late gestation à Necrosis

e.g. cerebral , cerebellar infraction , intestinal Atresia “ not duodenal ” **MCQ**

4- Dysplasia

Def Structural defect dt abnormal cellular organization or function, affect one general tissue type.

AE Mutant genes affect intercellular Metabolism .

e.g Osteogenesis imperfecta

Birth Defects

Types & AE :

Minor	Major	Single	Multiple
Cosmotic significance e.g. - clinodactyly - epican. fold - single in 14 % of newborn	If not corrected - impair body function ↓ life expectancy e.g. VSD Cleft lip Pyloric stenosis	- 2/3 of major cong Abn. - Affect only single body Site e.g. cleft lip cleft palate cong Ht Dse	Rule: 1 or 2 major + multiple minor

A/E of congenital anomalies

Syndromic multiple		Non syndromic single
Known AE	- Single gene mutation - Chromosomal Abn. - Environmental factors	- Single gene Mutation - Multi factorial (Gene + Env.) - Unknown
Unknown AE	- De Lange - Wiliam syndorme sporadic	

Genetics:

1- <u>Chromosomal 6%:</u>	AD	à	- Mental retardation - Growth retardation
	Sex linked	à	- less sever growth R. may not cause - Malformation.
2- <u>single gene 7.5%:</u>	Isolated	→	Hydrocephalus → X- linked Micophthalmia → AD/AR Ecterodactyly Poly " → AD
	Multiple "syndromic"		-Albert AD -EEC -Meckel AR -Robert
3- <u>Multifactorial:</u>	- majority of congenital Anomalies. - " usually isolated " non syndromic " - ASD , VSD - G.U : hypospedius , Renal agenesis - spina bifida - Other : cleft lip , palate		

4 - Environmental :

- * Drugs – infection – physical-
- * Recurrence Depend on Future Exposure to Tetratogens.

5 - **Unknown** 50% - single diaph. Hernia - anal atresia –Tracheo-eso fistula (low Recurre.)

Diagnostic approach to child with cong. Anomalies

A) History:

1- Family history:

birth defects , genetic diseases & multiple miscarriage.

2- Gestation:

duration , exposure to tetratogens & fetal movements.

3- Delivery:

complications of labor , fetal presentation , mode of delivery & neonatal status e.g. breathing & measurements.

4- Subsequent history:

physical growth (height, weight , H.C) & developmental progress (early milestones & formal psychometric testing)

B) Examination :

- 1- Child's behaviour, movements , facial expression & personality.
- 2- Detailed Anthropometric measurements.

C) Investigations:

1- Chromosome analysis:

from blood except skin biopsy in a stillborn baby .
+ fluorescent in situ hybridization (FISH)→ microdeletion syndromes.

2- DNA analysis.

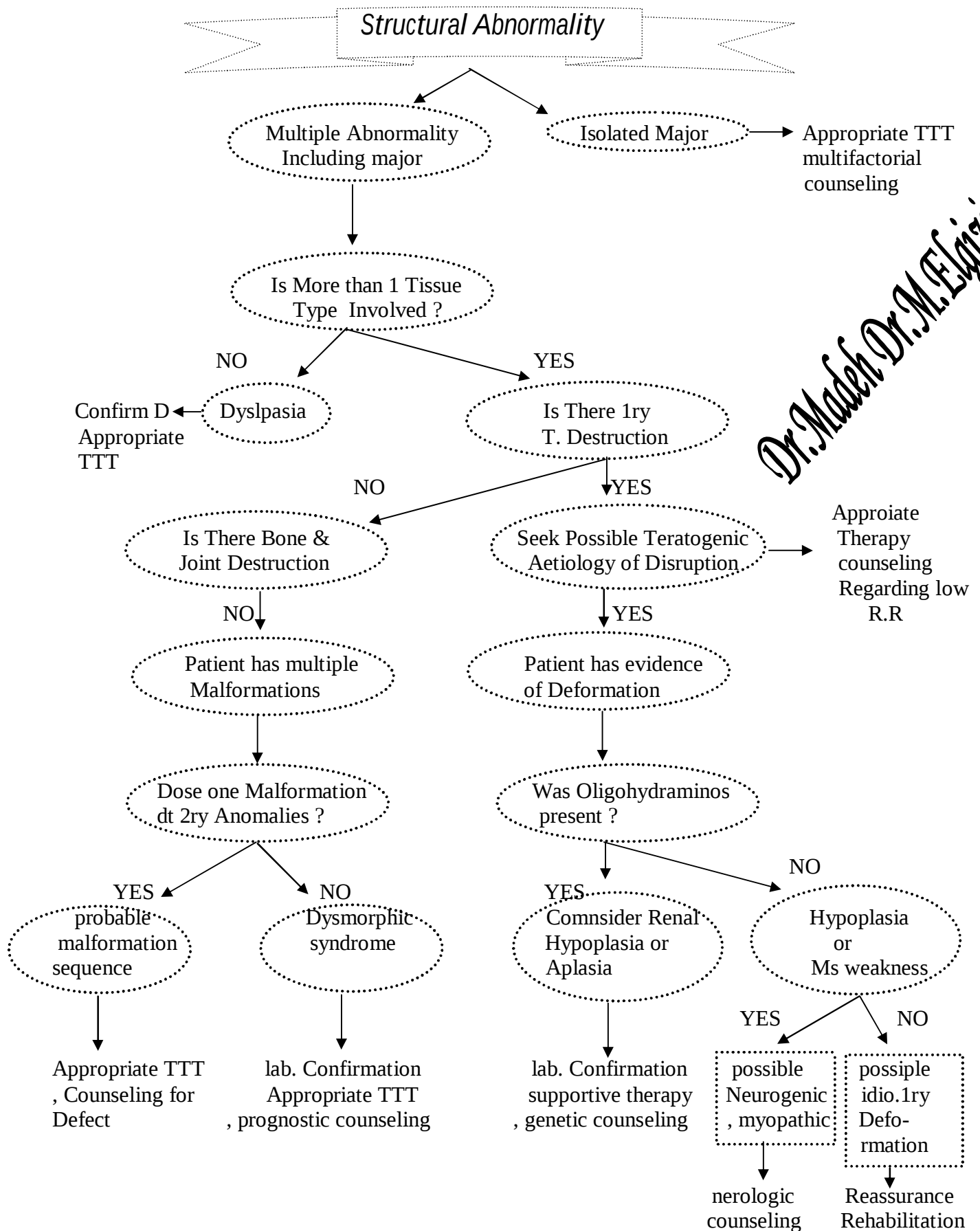
3- Radiographs U/S scans & other imaging techniques:

to detect heart & renal abnormalities.

4- Autopsy:

if the child has died to know the cause of death.

Dr.Madeh Dr.M.Elgizi



DOWN SYNDROME

Most common Autosomal Abnormality

- 1/ 660 هام شفوى وكليينيكال
- M = F

AE

1 - Complete trisomy 21(non-disjunction) الأشهر

- 95%
- 47 (xx +21)
- Non disjunction of chromosome 21 during meiotic division.
- Occur in oogenesis > spermatogenesis.
- Age Dependant , common in age >35 “ maternal age ”

2 - Mosaic D.S :

- 47(xx + 21 or 26)
- Non disjunctions of chromosome 21 early in division à 2 cell line (normal – trisomy)

3- Translocation : الأخطر

- 4%
- Most serious – young age < 25 (MCQ)
- 46 {xx t (14q +21q)}
- Chromosome 21 translocated to another group D " 14 " or gp G "26" à 45 Chromosomes but genotype 46 .
- Mother carrier à 2 possibilities à 4 types of gametes. شفوى
 - 1- normal
 - 2- Balanced Translocation Carrier
 - 3- Trisomy 14
 - 4- Trisomy 21
- Robertsonian i most common type.

Risk of recurrence :

- 1- Non- disjunction : if mother age 15 - 19 y` à 1 / 850 IF > 45 y à 1/50
- 2- Translocation : if 21/21 à 100%
- 3- mosaic : very rare

C/P

- M.R (never sever) شفوى
- *Development retardation*
- *Hypotonia*
- *Head* : Brachycephalic Microcephaly Flat occiput Silky hair.
- *Eye* :
 - Upward slanting
 - Hypertelorism " pseudo " MCQ
 - Medial epicanthic fold
 - Brush field spot

- Nystagmus
- Cataract
- Narrow palpebral fissure
- **Nose** : - Depressed nasal bridge.
- **Ear** : - small – low set – back . displaced.
- **Tongue** : - protruded - fissured
- **Neck** : - short - webbed
- **Hand** - brachydactyly - clinodactyly
- simian crease : Transverse palmar crease partial or complete
- **Foot** : - Wide space () big , 2nd toe
- Syndal line : deep planter crease

❁ Associated Cong. Anomalies ❁ شفوی

- 1) **Abdomen** : * Diastatic recti * Umbilical Hernia * Cong. Mega colon
- 2) **U.T** : * Imperforated anus. * Duodenal atresia (i commonest)
- 3) **CHD** : - 50%
 - Atreo-ventricular canal defect (commonest) هام
 - VSD

Complication :

- * **MR** : Accident (أشهر سبب لموته)
- * **GIT** : Anomalies
- * **CHD** : HF
 - Recurrent chest inf.(commonest Presentation)
- * **Leukemia** 20 times > G. population هام

Investigations :

Prenatal D	After birth
- chorionic villi sampling → 8wk - Amniocentesis → 16 wk - Mother Karyotyping in old age exposure to radiation	- karyotype : to Determine CytogenicType , Recurrence risk. - CVS : X- ray , ECG , Echo - BL : CBC à leukemia - Barium study à GIT anomaly - Radiology à U.T anomaly - Thyroid function Tests : (TSH ,T3 , T4)

<u>D.D</u>	Mongoloid facies	- Racial - Ch.hemolytic Anemia - congenital Hypertelorism	
	MR	- Cretinism - Microcephally - Hydrocephally	- Hydrocephalous C.P - Phenylketonuria - Galactosemia
	Other trisomy	18 Edward (Hypertonia), 13 Patou (polydactyly)	

TTT:

- Of cong. Anomalies & Associated disorder
- + Genetic counseling
- Social – Educational , Behavioral Therapy.

Prevention of genetic disorders

1) genetic counseling :

Def : Communication process estimate risk of developing of genetic disorder.

Goals : * to know mode of inheritance of genetic disorders

- * to know risk of recurrence .
- * concern parents whether their future child will be affected as their diseased child
- * cousins want to marry and worried about hazards of consanguinity
- * detection of heterozygous encourage them not to marry heterozygous .
- * on attempt to marry to concern about specific disease in the family.

Steps : * pedigree construction :

List patients relatives (age , sex , health status)
& refer to consanguinity .

- * Investigation according to nature of disease :
- * Estimation of risk of recurrence .

By placing the disease in one of four aetiological categories

- 1- Mutant genes.
- 2- Chromosomal aberration.
- 3- Multifactorial .
- 4- Environmental agents .

- * Take an action :

Parents decide the action either :

- 1- Prenatal diagnosis
- 2- Prevention of pregnancy

2) prenatal diagnosis:

Done by :

- * Chorionic villous sampling (CVS): at 10 weeks of gestation & under U/S guide.
- * Aminocentesis : primary diagnostic tool & at 16 – 20 weeks of gestation .
- * Foetal visualization :
 - a) U/S : - advantages : 1- no exposure to radiation

- 2- easy
- 3- guide amniocentesis.
- 4- detection of foetal abnormalities.

b) Fetoscopy : - Technique : fiberoptic telescope

- advantages : diagnosis of : 1- foetal malformation .
- 2- blood diseases.

c) Fetal radiography

d) Fetography : - injection of oil soluble dye into amniotic fluid → stick to vernix caseosa → define baby borders in X- ray .

e) Amniography : - injection of water soluble dye into amniotic fluid → contrast medium surround foetus in X-ray .

* Triple test :

- for : Down syndrome & chromosomal abnormality .
- by : Take an account : 1- maternal age .
- 2- biochemical markers :
 - α fetoprotein
 - unconjugated estradiol
 - HCG
- detect: 60 % of all Down syndrome .
- N.B. : شفوي الدكتور علي شلتوت :
new markers (Inhibin A) : glycoprotein secreted by placenta
Detect up to 80% of cases .

Indications :

- * Chromosomal abnormalities :1- previous child with chromosomal abnormality.
- 2- previous child with congenital anomalies
- 3- family with Down in second & third degree relatives.

3) Heterozygous " carrier " Detection :

- * Prevent disease by prevent marriage () heterozygous .

4) New born Screening :

- * For inborn error of metabolism
- * Early detection of cases :
 - specimen : dried blood on filter paper → phenyl ketonuria.
 - Dried urine // // // → amino aciduria

Developmental Abnormalities

Classification of Developmental disorders:

- Mental retardation . - Autism
- Attention deficit hyperactivity disorders - Language disorders
- learning disability - Visual or hearing impairment
- Cerebral palsy

Evaluation of Developmental Abnormalities

1- History

*** Warning signs:**

- Too floppy . - Too stiff - posturing . - Using one side more than the other
- Not socially interacting. - Not doing what other infants in his age are doing
- Not talking

*** Family history :**

- Consanguinity . - Neurological difficulties -- Psychiatric difficulties
- Social difficulties

*** Prenatal history:**

- Teratogenic exposure . - Infectious illness - Addictive substance -- Trauma.

*** Perinatal history:**

- Gestational age . - Birth wt . - Apgar score . - Medical complications

*** Past history:**

- Ch. Resp. illness . - Recurrent OM . - Head trauma . - Obstructive sleep apnea

2- Physical examination:

- o **Growth parameters** : - wt, height, head circumference
- o **Dysmorphic features.**
- o **Signs of neuro-cutaneous disorders** e.g. Neurofibromatosis.
- o **Eye** : cataract in Galactosemia

3- Investigations:

1- **Laboratory tests** : i- Thyroid function tests

ii- Lab. tests for inborn errors of metabolism . iii- Serum lead

2- **Electroencephalography (EEG)**

Not routinely indicated . Only if there is seizures encephalopathy , Microcephaly
macrocephaly

3- **Neuro imaging** : - Brain MRI is more sensitive than CT

4- **Vision & hearing assessment.**

5- **Chromosomal & Cytogenetic studies.**

Management of Developmental delayed child

- 1- Initial diagnostic evaluation
- 2- Early intervention program (Physiotherapy . Speech therapy)
- 3- Identify & treat associated problems : Vision , hearing, seizures.....ct.
- 4- Orthopedic .
- 5- Psychological support the parents

*Teratogenic agents & their Effects on child health**Def of Teratogen :*

Any agent that can produce malformation (birth space) by interfering with normal embryonic development

Difference () teratogen & mutagen:

- Teratogen acts on the somatic cells of the developing embryo & produces birth defects
- Mutagen acts on germ cells & alter the genetic material

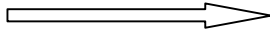
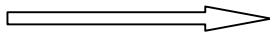
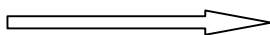
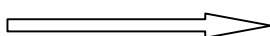
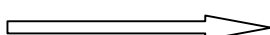
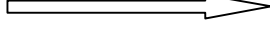
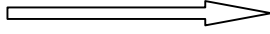
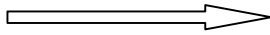
Factors affecting teratogenicity:

- 1) Time of exposure
- 2) Dosage.
- 3) Hereditary predisposition
- 4) Specificity of teratogens: Certain teratogens cause characteristic patterns of malformation

Classification of teratogenic agents.

- 1) Drugs & chemicals
- 2) Maternal infections - viruses bacteria & parasites
- 3) Physical agents : - Ionizing radiation - Prolonged hyperthermia
- 4) Maternal diseases : - Maternal DM . - Maternal phenylketonuria

I- Common Teratogenic drugs:

<i>Drug</i>		<i>Effects</i>
Alcohol		Fetal alcohol syndrome
Lithium		Ebstein anomaly
Phenytoin		Cardiac_defects ,cleft palate , hypoplastic nails
Chloroquine		Chorioretinitis , Deafness
Warfarin		Nasal hypoplasia Stippled epiphyses
Streptomycin		Deafness
Tetracycline		Enamel hypoplasia
Valproic acid		Neural tube defects

Fetal alcohol syndrome

- Growth retardation (prenatal & postnatal)
- Microcephaly
- Developmental delay.
- Skeletal & cardiac anomalies
- Characteristic facies : (mid face hypoplasia)
 - * Short palpebral fissure
 - * Epicanthic folds
 - * Short upturned nose
 - * Thin upper lip

II- Maternal infections:.

Infectious Teratogenic Agents :

<i>Infection</i>	<i>Effect</i>
Viruses:	
CMV	Microcephaly - chorioretinitis
Hereps Simplex	Microcephaly - retinitis
Varicella Zoster	Microcephaly - chorioretinitis
Bacteria :	
Syphilis	Hydrocephaly - chorioretinitis
Parasites:	
Toxoplasmosis	Microcephaly - chorioretinitis - retinitis

congenital Rubella syndrome:

- * Cardiac : - Pulmonary artery stenosis (55 %) - Patent ductus arteriousus (43 %)
- * Deafness
- * Cataracts, Glaucoma, retinopathy.
- * Mental retardation
- * IUGR.
- * Neonatal purpura , hepatosplenomegaly.

*III- Physical agents:**Ionizing radiation:*

- © Heavy doses of ionizing radiation : - Microcephaly - Occular defects
- © The most sensitive time of exposure is from 2- 5 weeks after conception
- © Risks of low dose diagnostic procedures are minimal.

Prolonged hyperthermia:

© Prolonged hyperthermia in early pregnancy : - Microcephaly , Microphthalmia, Encephalocele.

*IV- Maternal diseases:*Maternal D.M:

* Most common malformation in infants of diabetic mother include:

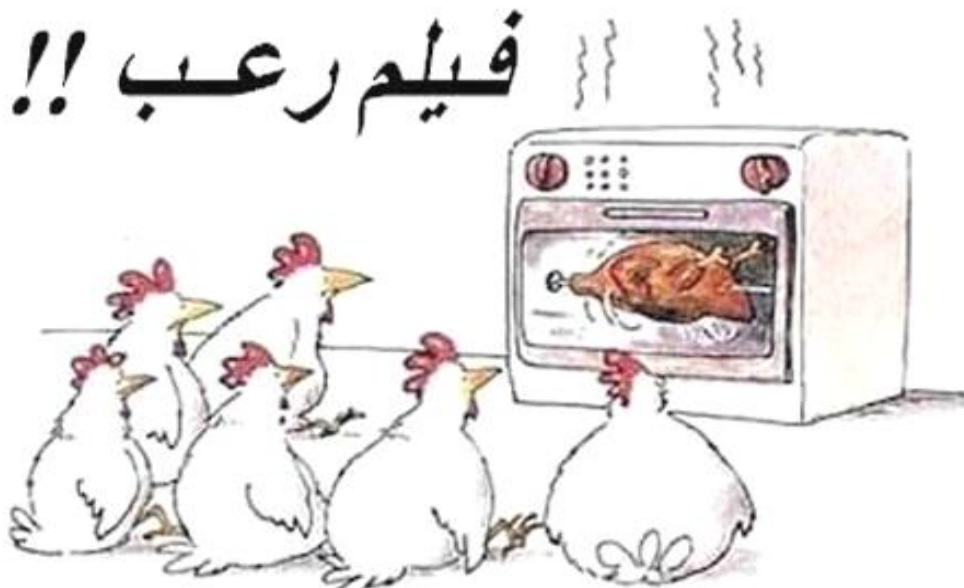
- Congenital heart disease .
- Lumbo-sacral dysgenesis .
- Renal disorders :
 - Double ureter .
 - Renal vein thrombosis .
 - Renal agenesis
- Neural tube defects
- Small lt. colon syndrome

Maternal phenylketonuria:

* High level of phenylalanine in pregnant woman with PKU not on special diet

→ serious damage to the fetus :

- Mental retardation (100 %)
- Congenital heart defects
- Microcephaly



Dr.Madeh *** Dr.AlgizY

Rickets

Def. Its a systemic metabolic dis. Charact. Pathologically by defective bone mineralization & clinically by various skeletal , muscular & neurological manif.

Types (Classification of Rickets) نظري

I- Vit. D deficiency rickets:

1- Congenital rickets :

- mother has osteomalacia.
- Main presentation → hypocalcaemia & tetany after birth.

2- Infantile rickets → commonest type. MCQ. (3m – 2 yrs)

3- Late infantile rickets → rare (5 - 15).

4- Atrophic rickets (ch. By osteoporosis) → associated with PCM (no manifestations → diagnosed by lab. Investigation).

5- Subclinical rickets → no manifestation but diagnosed by X- ray & lab. Investigations.

II- Vit. D resistant rickets

a- 1 ry type :

- 1- Hypophosphatemic. (pH^+)
- 2- Hypocalcaemia (vit. D dependant rickets).

b- 2ry type :

- 1- Renal type.
- 2- Hepatic type.
- 3- Onchogenic hypophosphatemia with Mg.
- 4- Iatrogenic "drug induced " e.g. anticonvulsants.
- 5- Malabsorption " celiac ".

III- Related cond. Resembling rickets

- 1- Hypophosphatasia.
- 2- Hyperparathyroidism.

N.B

1. Rickets : Inadeq. Mineralization.
2. Osteomalacia: failure of mineralization.
3. Osteoporosis : Demineralization.

*** Vit. D :**

Nature : Its a fat soluble vit.

Types :

	Vit.D2 " Calciferol"	Vit.D3 " Cholecalciferol"
Origin:	Plant	Animal
Produced by:	Irradiation of plant Ergosterol	Irradiation of skin 7-dehydro- cholesterol

Actions : (by 1,25 DHCC)

1. On intestine : regulation of Ca & phosphate absorption.
2. On the bone : helps mineralization of bone matrix.
3. On kidney : regulation of Ca & phosphate reabsorption by the renal tubules.

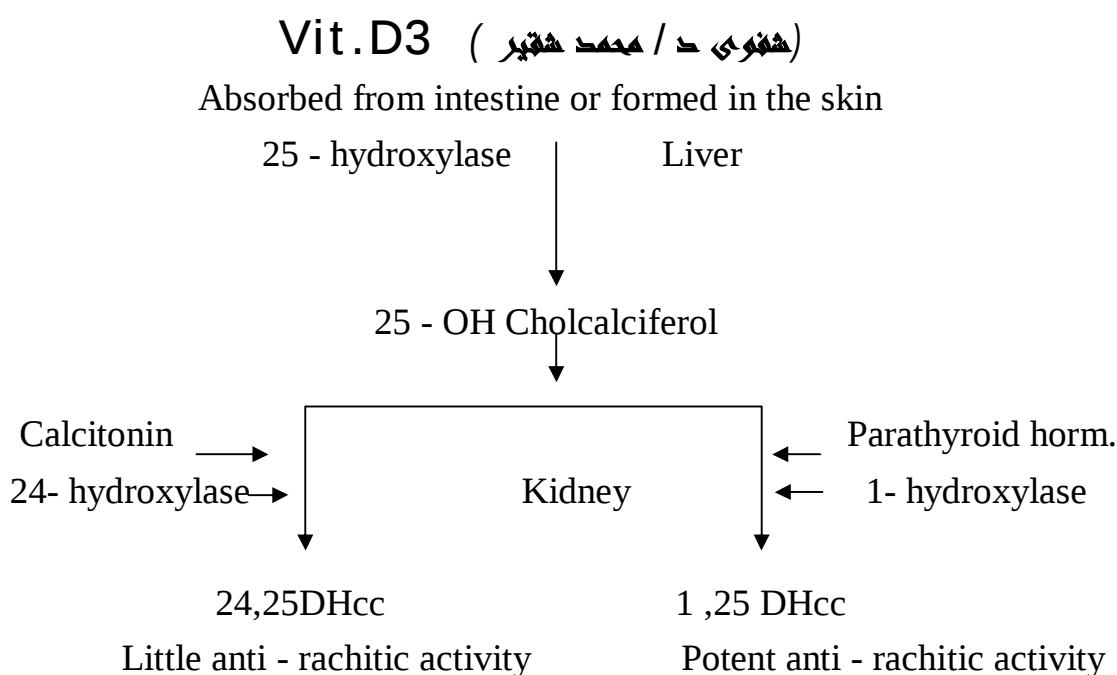
Sources

1. Foods rich in Vit.D (fish liver oil . egg yolk & fortified dried milk).
2. Foods poor in Vit.D (breast milk , animal milk & CHO).

Metabolism

It is absorbed from the intestine in the presence of bile.

Carried by a2 globulin to be stored in the liver.



Deficiency of vit. D leads to :

- | | |
|----------------------|---------------------|
| 1- Infantile rickets | 3- Infantile tetany |
| 2- Osteomalacia. | 4- Dental decay |

Infantile Rickets

* Incidence:

- Age : () 3 months -2 years.
- Season : more common during Winter.
- Race : more common in Negros.

* Etiology

1- Dietary causes "Rachitogenic diet" : (\$intake)

- The rachitogenic effect of diet may be due to any of the following:
 - 1- Low content of vit.D e.g. breast milk, cow's milk & CHO diet.
 - 2- High content of vit. D antagonists (phytic acid) e.g. cereals & veget.
 - 3- Low content of Ca & phosph. e.g. CHO (1.5: 1 is the best for absorption)
 - 4- Unsuitable Ca / Phosph. ratio e.g. cow's milk
 - 5- High Content of Ca chelating agents e.g. oxalates & citrates.
 - 6- Alkalinity which will##PH of G.I.T. & thus \$ absorption of vit.D.

2- Impaired absorption of vit.D: (\$ absorption)

- a- Fat intolerance. b- Ch. Recurrent diarrhea. c- Coeliac disease.
- d- Steatorrhea. e- Cystic fibrosis. f- Ch. Hepatic disease.
- g- Drugs e.g. anticonvulsants (phenob.).

3-Failure of activation of vit.D in superficial layers of skin: (\$activation)

- Due to lack of exposure to the sun rays.

4- Congenital rickets: (congenital)

- Occurs in breast fed infants borne to mothers suffering from severe osteomalacia

*Pathology of active Rickets: Main pathological features are :

1- Abnormal proliferation of cartilage: due to defective mineralization :

At various sites of body leading to:

- 1- Enlargement of epiphyseal ends of long bones.
- 2- Swelling of costo-chondral junctions.
- 3- Bossing of frontal & parietal eminences of skull

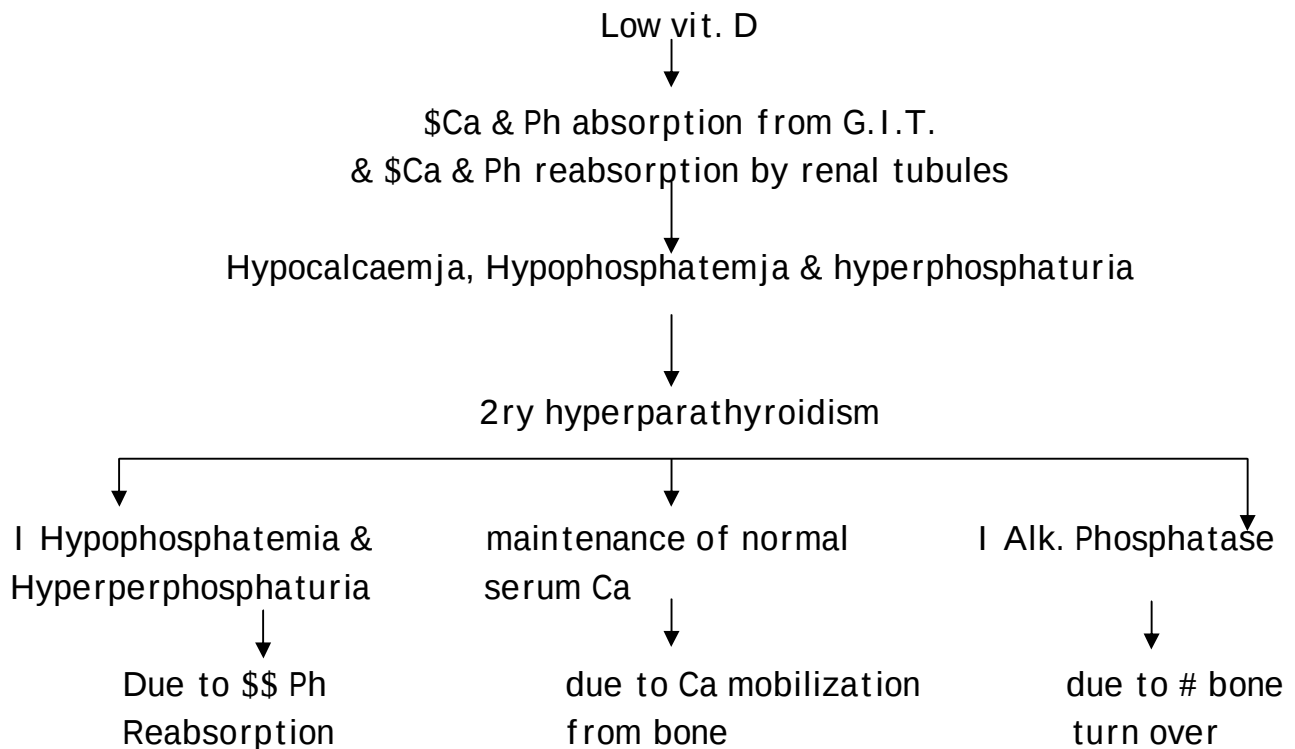
2- Irregular invasion of metaphyseal area:

By abnormal layers of proliferating cartilage more centrally than Peripherally leading to typical cupping & fraying seen in X- ray films of long bones at metaphyseal area.

3-Irregular subperiosteal deposition of osteoid tissue:

Leading to irregularity & thickening of cortex. (double periosteal line)

Pathogenesis : = Pathogenesis of Biochemical changes



* Some exception . MCQ & شفوي

1- Serum Ca may be \$: causes of hypocalcemia in infantile Rickets = causes of tetany in inf. Rickets

1. Failure of 2ry hyperparathyroidism
2. In late stages with depletion of bone Ca.
3. After shock therapy.
4. As a complication for G.E. due to \$absorption from intestine.
5. Exhausted parathyroid.

2- Serum alkaline phosphatase : may be normal if it's associated with protein depletion (atrophic rickets). KWO

C/P :

A- Manifestations of early rickets

I- Biochemical change: Especially high alkaline phosphatase levels are earliest manifest. of rickets.

II- Radiological changes: They occur before appearance of clinical manifestations.

III- Clinical manifestations:

- a- Symptoms: 1- Excess head sweating. 2- Irritability. 3- Anorexia.
4- Constipation.

* May be due to dysfunction of the ANS.

b- Signs :

1- Craniotables:

- i- It's a ping pong feeling of skull bones when compressed by fingers indicating their softness & thinness
- ii- It tends to be localized over occipital & post. Part of parietal bones
- iii- Rarely presents before 3 months of age & disappear. Before the end of the 1st year

2- Rosary Chest.

B- Manifestations of advanced rickets:

§ Symptoms : 1- Delayed sitting, standing , walking & teething.
2 - Bone deformities.

§ Signs : delayed mile stones

I- Skeletal manifestations

1-Head:

1. Enlargement & symmetry.
2. Frontal & parietal bossing.
3. Box - shaped head or caput quadratum.
4. Depression at root of nose.
5. Delayed closure of ant. Fontanell.
6. Delayed eruption of teeth & tends to decay.

2- Chest:

- 1- Rosary beads : a- these are small palpable swellings
b-At costochondral junctions sparing 1st & 2nd ribs.
c- They are not mobile.
- 2- Harrison's sulcus :
a- It's a transverse groove at lower chest wall at costal insertion of diaphragm.
b- Produced by dragging effect of diaph. During resp. movem. on soft bones of chest.
- 3- Longitudinal sulcus :
- It's a vertical groove behind rosary beads resulting from compression of ribs by atmospheric pressure at the weakest points
- 4- Pigeon chest: - It's result of protrusion of sternum.
- 5- Flaring of lower ribs: with eversion of costal margins.

N.B: Consequences of chest deformities:

- 1- Limitation of chest capacity for expansion susceptibility to chest infection, atelectasis.
- 2- Spleen & liver may be palpable due to downward displacement by deformed thorax.

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3-Abdomen:

- 1- Abd. Protrusion is nearly a constant finding in infantile rickets.
- 2- Palpable liver & spleen.
- 3- Umbilical hernia.

4- Pelvis (*Rachitic pelvis*):

- 1- Contracted inlet : due to forward projection of promontory of sacrum produced by exaggeration of lumbar lordosis.
- 2- Outlet contraction: due to forward projection of tip of coccyx.

5- Extremities:

- 1- Enlargement of ends of long bones esp. at wrist & ankle.
- 2- Pathological fracture" green stick fracture→ painful.
- 3- Marfan's sign:
It's a transverse groove felt by palpation at lower ends of long bones mostly over tibia just above medial malleolus giving impression of double epiphy. Due to abnormal proliferation of cartilage at 2 different ossific centres instead of one → producing 2 elevations with a groove in ()
- 4- Deformities of long bones :
It affects upper more than lower limbs in crawling infants & vice versa in standing & walking infants.
- 5- Deformities of joints : due to hypotonia of ms & laxity of ligaments
e.g. genu varum & genu valgum.
- 6- Spine: Kyphosis , scoliosis & kyphoscoliosis with compensatory exaggeration of lumber lordosis are often present

II - Muscular manifestations:

Hypotonia may be due to hypophosphatemia
(flabby infant , pot belly abdomen - frog like , constipation)

III - Neurological manifestations:

- 1- Early : in the form of sweating, anorexia, irritability & constipation.
- 2- Late : in the form of tetany.

Investigations

A- Radiology of Rickets:

- X- ray of the lower end of the radius & ulna bec. :
 - 1- Readily accessible.
 - 2- Site of rapid growth.
 - 3- The bone are small & The surrounding tissues are small.

	<i>Active rickets</i>	<i>Healing rickets</i>	<i>Healed rickets</i>
End of long bones (epiphysis & metaphysis)	- Widened (broadening) - Cupped (concavity) - Frayed: due to: loss or haziness of line of provisional calcification. Triad of active Rickets	- Widened (broadening) - Cupped (concavity) - No fraying - Faint narrow irregular band (due to mineralization of provisional zone of calcification with ttt) it will appear () 2 -3 wks	- No -No. -No. Continuous line of Thick density.
Distance () lower end of radius & ulna & carpal centre of ossification	# due to large ricketic metaphysis which not calcified.	## due to large ricketic metaphysis which not calcified.	Normal
Diaphysis (Shaft) * Density: * Periosteum : * Path. Fractures : *Deformity:	* \$(Rarefaction). * Double periosteal line dt subperiosteal deposition of osteoid tissue which is translucent) * May be present. * May be present.	* \$(Rarefaction). * Double periosteal line (due to subperiosteal deposition of osteoid tissue which is translucent) * May be present. * May be present.	* Normal. * No * Healed, * May present.

N.B : In healed type : Continues line of thick density & deformity used to D.D.
 X- ray of healed rickets from that of normal bone.

B- Lab. Investigations (Biochemical changes of infantile rickets) :

I - Blood changes

1. Serum Ca: normal (9-11 mg/100 m).
2. Serum phosphorus \$ (normal 4.5 - 6.5 mg / ml).
3. Serum alkaline phosphatase # (normal 5 - 15 Bodansky units) - most valuable test for detection of early rickets.
4. Serum 1,25 DHcc \$ (normally 36 pg/ml).

II - Urine changes:

1. Absent Ca: no calcuria.
2. Hyperphosphaturia

D.D. of Rickets:I- Causes of delayed walking : **See neurology**

II- Causes of Craniotables:

- | | |
|-----------------------------|-------------------|
| 1- Rickets. | 2- Hydrocephalus. |
| 3- Osteogenesis imperfecta. | 4- Prematurity. |
| 5- Hypervitaminosis A. | |

III- Causes of depressed bridge of nose:

- | | | |
|--------------|--------------------------|-------------------|
| 1- Rickets. | 2- Mongolism. | 3- Cretinism. |
| 4- Familial. | 5- Ch. Hemolytic anemia. | 6- Hydrocephalus. |

IV- Causes of rosary chest:

- 1- Rickets (rounded due to rack. Metaph.).
- 2- Marasmus
- 3- Scurvy (angular due to sublaxation of sternal & costal parts of ribs).
- 4- Chonrodystrophy

V- Causes of delayed Closure of ant. Fontanel.:

- | | | |
|---------------|----------------------------|-----------------|
| 1- Rickets. | 2- Hydrocephalus. | 3- Mongolism. |
| 4- Cretinism. | 5- Osteogenesis imperfecta | 6- Prematurity. |

Complication of Rickets:

1- Respiratory complication..

- Chest infections with or without atelactasis are frequent & are due to
 1. Chest deformity → \$expansion & vital capacity.
 2. Hypotonia of resp. ms. With weakness of the cough reflex.

2- G.I. T. complication:

- 1- Alternating diarrhoea & constipation due to hypotonia of intestinal & abd. Ms.
- 2- 2ry bacterial infection.

3- Others:

- 1- Anemia: due to iron \$ or associated 2iy infection.
- 2- Tetany.
- 3- Skeletal deformities
- 4- Dwarfism.
- 5-. Complications of therapy.
- 6- Obstructed labour in Racketic mother.

Treatment of infantile rickets:

I Prophylactic ttt:

1. Exposure to sun rays.
2. Prophylactic administration of vit.D (in normal infant 400 - 800 IU/ d from 2nd month but in preterm 1000- 1500 IU / d from 1st month.)
3. Administration of vit.D to pregnant or lactating mother.
4. Vit.C 25 mg/d (as vit.C is acidic which help solubility & #absorption of Ca).

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II- Curative ttt:

A- Specific therapy:

- Vit.D3 : * *Orally* :1500- 5000 IU / day for 6-8 weeks.
- * *I.M* : *single 600,000 IU/ month , max. 3 months.
- * S.E.: Shock therapy Ca shift from blood to bone -tetany.
- * Maintenance : 400 - 800 IU / day orally.

B- Adjuvant therapy:

- Adequate Ca & phosphorus intake.
- Vit. esp. vit. B complex, vit. C & iron.
- Exposure to ultraviolet sun rays better from 5 - 8 a.m. & 3 - 5 p.m.

C- Actinotherapy : UVR obtained from special Hg. Lamp.D- Prevention of bones deformities : by avoiding walking.E- TTT of complications:

1. Tetany : Ca gluconate 10 % I.V.I.
2. Bone deformities: osteotomy.
3. TTT of infections e.g. pneumonia.

Non – vit. D deficiency rickets " Vit. D resistant rickets)

	Vit. D resistant hypophosphatemic rickets	Vit. D resistant hypocalcaemic rickets
Aetiology	* Inability of the kidney to reabsorp. Ph. Due to defective hydroxylation of vit. D in the kidney .	* Type I : inability of the kidney to reabsorp. The Ca. * Type II: endogenous resistance to Ca.
Genetics	* X- linked dominant more in ♀ > ♂	* A.R.
C / p	- Bone deformity esp. in the L.L. - \$linear growth. - No affection of general health - No hypotonia. - No head & chest involvement	As VDDR
Lab	* \$Ph in serum. * #Ph in urine.	* \$Serum Ca. * Normal Ph.
TTT	* Vit D massive dose * Phospaate infancy : $\frac{1}{2}$ - 1.5 gm / d after inf : 1.5 – 3 gm / d * Osteotomy	* Vit D Massive dose Osteotomy

Def : Rickets resistant to the usual dose of vit. D.

Types:

(1) Renal rickets:

1- Renal glomerular rickets:

A/E : C.R.F.

Pathophysiology:

- * ph. Retention → hyperphosphatemia → ↓ Sr. Ca^{++} →
2ry hyperparathyroidism & bone resorption .
- * Failure of vit. D activation.

2- Renal tubular rickets:

Isolated defects:

- * Vit. D resistant hypophosphatemia rickets.
 - X-linked dominant disorder.
 - Failure of renal tubules to reabsorb ph.
- * Vit. D resistant hypocalcemic rickets.
 - Autosomal recessive.
 - Failure of renal tubules to reabsorb Ca^{++} .

Multiple defects:

1. Fanconi syndrome:

Proximal tubular defect characterized by (phosphaturia – glucosuria – aminoaciduria – ↑ excretion of K , Uric acid , Na.

2. Lignac syndrome : fanconi + cystinosis .

- 3. Lowe syndrome:**
- Oculo-cerebro renal syndrome
 - Ocular → glaucoma – cataract.
 - Cerebral → M.R.
 - Renal → like fanconi

4. Lightwood syndrome:

- * Inability to form acidic urine due to loss of HCO_3

* Metabolic acidosis & hypophatemia

(2) Hepatic rickets:

Chronic liver dise.: defective activation & absorption of vit. D.

(3) Hyperparathyroidism.

(4) Congenital hypophosphatasia:

Congenital absence of alk. Phosphatase → defective mineralization of bone

→ rickets & teeth loss.

ttt: cortisone (no value of vit. D).

Classification of PEM

1- Wellcome classification: body wt $\pm$ edema.

* $> 80\%$ of expected wt for age → normal.

* $60-80\%$ of exp. wt for age → - With edema → KWO.

- Without edema → simple undernutrition.

* $< 60\%$ of exp. wt for age → - With edema → MKWO

- Without edema → marasmus.

2- Jellife classification:

- Mild or moderate undernutrition.
- Nutritional dwarfism.
- KWO.
- Marasmus.

3- Waterlaw classification (WHO recommendation).

Height for age: measure stunting → index for chronic malnutrition.

Wt. for age: measure wasting → index for acute malnutrition.

K.W.O. (red body)

(Acute disease not chronic)

Def: Clinical syndrome d. to inadequate supply of protein & a.a.

Incidence:

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- Age (6m- 2yrs.).
- Not before 6m as breast milk is sufficient.
- Not after 2 yrs. As body get mixed food.
- Disease starts as weaning begins as the new diet is composed mainly of CHO & low protein , a.a.

C/P:

a- Constant manifestations: present in all cases:

- 1- Growth retardation & Ms. Wasting.
- 2- Edema.
- 3- Psychomotor changes.

b- Variable manifestations: not present in every case:

- | | |
|-------------------|------------------------------|
| 1- Skin changes. | 4- Anaemia & Vit deficiency. |
| 2- Hair changes. | 5- Infection. |
| 3- GIT disorders. | 6- Trace elements. |
| 4- Hepatomegally. | 7- Anthropometric measutes. |

(A) Constant features :

1- Growth retardation & Ms. Wasting:

- * Wt.: 1s fails to gain wt. then progressive loss of wt (60-80%).
- * Height & Head circumf: less affected.
- * Subcutaneous fat: preserved.

- * Muscle: thin, weak ↓ mid arm circumference (< 12.5).

2- Oedema:

- * Start in face & eye lid → hands → generalized.
- * Never ascites & Pl. effusion.
- * D. to: 1- Hypoproteinemia (↓ intake, absorprion, ↑ loss).
2- ↑ ADH & aldoserone.

3- Psychomotor changes:

- * Leathergy
- * Loss of interest.
- * Miserable look.
- d.t : * ↓ Mn. * Nicotinic acid .
- * Aromatic a.a. * Thyroid function

(B) Variable features :

1- Skin changes:

- * Dermatitis (common in flexure sites).
- * Etythema → hyperpigmentation → dequamation → depigmentation → ulceration.
- * d. to: ↓ * Vit. A. * Nicotinic acid.
- * Suprarenal function * Essential fatty acids.

2- Hair changes:

- * Lusterless, curileness, fine, easily pickable.
- * Colour changes (Reddish - grey or white in severe cases).
- * Discoloration is segmental giving flag sign (pale zones sepatated by pigmented zone).
- * Due to: * ↓ (Cu - Sulphur containing a.a –Tyrosine- Pantothenic acid)

3- GIT disorders:

- * A,N,V, D., steatorrhea (d. to 2ry infection).
- * d. to: infection inside & outside the gut.
- * ↓ Intestinal & pancreatic enzymes.
- * ↓ Intestinal disaccharides → fermentive diarrhea.

4- *Hepatomegally*: (No cirrhosis only if toxic viral hepatitis).

- * d. to: - ↑ Flux of F.A (fatty liver). - ↓ Hepatic synthesis of B-lipoproteins.

5- *Anaemia & vit. Deficiency*:

- * Microcytic → Fe ↓ .
- * Normocytic normo. → d. to infection, protein deficiency.
- * Macrocytic → d. to ↓ vit. B12 , Folic acid.
- * Hemorrhagic d.to ↓vit. K., ↓ liver function, ↓ prothrombin
↑ Capillary permeability.

6- *Liability to infection esp. T.B. so x-ray important.*

7- *Trace elements*: - ↓ Fe : anemia.

- ↓ Mn : psychomotor dise.
- ↓ Zinc: growth retardation.
- ↓ Cu : hair changes.

8- *Anthropometric Measures*: as before.

Complications:

- Recurrent inf. (T.B. , HIV) - Gastro-entritis.
- Hypo glycemia. - Hypo thermia.
- HF

N.B. : Most common cause of death → Chest infection.

Investigations:

Subclinical cases:

- Total s. protein → normal.
- Albumin → ↓
- Urea N2/ creatinine N2 ratio → (8-12)
- Non-essential a. a./ essential a. a. ($< 2 \rightarrow$ normal , $2-3 \rightarrow$ pre-Kwo, $> 3 \rightarrow$ KWO).

Clinical cases:

- ↓ Albumin, ceruloplasmin, transferrin.
- Total a.a. → normal or ↑.
- Essential a.a. → ↓.
- Non- essential a.a → ↑ except (arginine – citrullin).
- Non-ess. / essent. a.a. > 3
- ↓ Serum glucose & flat glucose curve.
- ↓ Serum & urine urea.
- ↓ Serum enzymes.
- ↓ Serum Fe , Cu , Mg , vitamins.
- ↓ Cholesterol, triglycerides.
- Creatinine excretion.
- Anemia :.....
- X-ray chest → T.B.

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1- Prophylactic ttt:

- * Encourage breast feeding with supplementation.
- * Proper weaning on high protein & balanced diet.
- * Immunization against infectious disease.
- * Early detection of malnutrition.

*2- Curative ttt:***a- Hospitalization.**

b- TTT of the cause.

c- Dietetic ttt: * to ↑ protein intake gradually to 4-6 gm/kg/day)

* If the patient:

<i>Weaned</i>	<i>Not weaned</i>
Diet rich in protein Youghourt: • Acidified. • Rich in protein. • Thick. • Can be skimmed.	• Start → skimmed acidified milk (↓fat – ↑protein – easy digestible) • Then → half cream milk. • If tolerated → full cream or protein milk. • If lactose intol. → Lactose free formula. • Then → diet rich in protein • Amount → start 90 ml/kg/day → ↑ 150 ml/kg/d

Blood transfusion:

- ↑ Hb. for Anaemia.
- ↑ Plasma for protein.
- ↑ Antibodies for inf.
- ↑ Enzymes for anorexia.
- ↑ Clotting factors for bleeding.

Vitamin supplement:.....**d- Complications ttt:**

- * Infection : antibiotics.
- * Anemia : (bl. Transf., Iron, vit. B12 & folic acid)
- * Diarrhea : ttt of dehydration – lactose free diet.
- * Xerophthalmia: vit. A drops , Antibodies , atropine ointment .
- * Hypocalcaemia : IV or oral Ca.

Recovery of KWO: * Smile : 4 days. * Edema : 10 days.

* Complete : 1-3 months * Death : 15%.

Marasmus = infantile atrophy

Def: Chronic state of undernutrition characterized by progressive wt. Loss
(< 60% of exp. Wt. for age).

A/E:

B Prematurity: - Unable to suckle well

- ↓ digestion & absorption.

- ↑ rate of growth.

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C Congenital anomalies:

* MR

* CHD

* C. Pyloric stenosis

* Liver cirrhosis

* Cong. Anomalies of U.T.

D Diet:

- ↓ intake of calories:

* **Quantitative:** - Scanty milk supply.

- Insuff. Amount of milk.

- Insuff. Caloric intake .

* **Qualitative:** - Prolonged by feeding.

- Formula (sugar – very diluted)

- Fluids after gastroenteritis.

* **Feeding difficulties:** - Mechanical : cleft palate

- M.R.

- ↓ Diagection: cystic fibrosis.

- ↓ Absorption: malabsorption syndrome.

- ↑ loss of food substances : vomiting & diarrhea.

E Enviyonmental: poor classes with poverty.

Endocrinal : * DM * Hyperthyroidism * Addison

i Infection : * T.B. (chronic or repeated acute).

- M Metabolic** : * CHO : glycogen storage disease
* Fat : gaucher disease.
* Electrolytes: infantile renal tubular Acidosis.

C/P : Insidious onset of:

1- Growth retardation & Ms. Wasting:

- * Wt.: progressive loss of wt < 60%.
- * fat: S.C. fat is lost:
 - Skin: wrinkled, loss elasticity.
 - Face: senile appearance (3rd degree).
 - Abdomen: scaphoid or distended.
 - Limbs: thin sticks.
- * Ms.: Wasting, thin atrophic, ↓ mid arm circumference.
- * Ht , head, chest, abd. Circumference: less affected.

- 2- GIT:** * Constipation common.
* Gastroenteritis.
* Starvation stool (small dark).

3- Liver & spleen : small size.

4- Anaemia & vit. Deficiency.

- 5- CVS:** * Peripheral circulation failure.
* Weak heart sound.

- 6- Vital signs:** * Pulse: slow feeble.
* B.P. ↓
* Temp: subnormal with cold & blue extremities.
* Respiration: shallow.

7- Recurrent infection: Most common: - Bronchopneumonia
- Gastroenteritis.

Degrees of marasmus:

	<i>1st</i>	<i>2nd</i>	<i>3rd</i>
<i>Wt. Loss</i>	40%	41-49%	> 50%
<i>S.C. fat</i>	Over the abdomen	Over the buttock & extremities	Face & loss buccal Bad of fat (lastly due to Unsaturated F.A.)

Complications:

- * Recurrent infection. * Gastroenteritis
- * Glycemia * Thermia (bad prognosis)
- + : * Atrophic ulcers: over pressure sites.
- * Purpura: bad prognosis.
- * Oedema: MKWO.

Prognosis : defend on:

- * Degree * Complication: present or not.
- * Cause: correctable or not. * Age : worse if young.

Investigations:

- Plasma proteins: ↓ but not as KWO.
- Glucose ↓ & flat sugar curve
- Anemia - leukocytosis - starvation Ketonuria

TTT:

a- Prophylactic : as KWO

b- TTT of the cause : as KWO

c- Dietetic ttt:

- Diet:** - ↑ caloric intake gradually up to 150-200 cal/kg/d:
- Gradual ↑ of food intake.

- Amount: calculated acc.to actual wt. Then ↑ gradually till reach Amount for age.
- In older infant protein food recommended.
- Fluids in () feeds d. to dehydration & constipation.
- Milk formula:
 - 1st lactose free then, skimmed acidified.
 - Then, 1/2 cream milk.
 - Then, full cream milk.

Blood transfusion : as KWO

Vit. Supplementation : as KWO

d- ttt of complications : as KWO

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Vit. A ⁻⁻⁻	Vit. C ⁻⁻⁻ (scurvey)
A/E:	A/E:
C/P: (a) <u>Ocular manifestations:</u> 1- Dry eye. (xerophthalemia: 1 st sign) 2- Night blindness 3- Keratomalacia 4- Panophthalmitis 5- Bitot's spots (b) <u>Extra-ocular:</u> - Skin: dry , hyperkeratosis - Infection : ↑ - ICT: benign ↑ - Growth: ↓ physical & mental	C/P: Appear after 3-5 m after prolonged deficiency * <u>Apprehensive child & irritable:</u> tender painful limbs d.to hemorrhage under peritoneum. * <u>Bleeding</u> * + <u>Rosary chest</u> * <u>Pain:</u> pseudoparalysis, frog like position
TTT: Vit A. Orally or IM 5000-25000 U/d + Diet.	TTT: Vit. C orally or IV 100-500 mg/day + Diet (orange juice) If ↑↑ dose: - GIT irritation – diarrhea - Oxalate stones.
Vit. A - -	Vit D - -
Acute: N, V, drowsiness, ↑ICT benign Chronic: - Anorexia - alopecia - Pruritis – bone tender swelling - Craniotabes - Liver: enlarged - CNS: ++ irritability	Causes: ↑ Intake: - big dose - anxious ptn – concentrated form C/P: - Anorexia – pallor - Hypercalcemia– hypercalcuria - Renal failure – hypotonia - Polyuria – polydipsia - dehydration – constipation
X-ray: Hyperostosis of long bone	X-ray: * Metastatic calcification * Dense metaphyseal line * ↓ density of shaft
TTT: stop vit. A	TTT: - Stop Vit D & give cortisone - ttt of Ca (AlOH3) - Symptomatic ttt .